



INDIAN JOURNAL OF PRACTICAL PEDIATRICS



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SUPPORTIVE AND PALLIATIVE CARE

INTRODUCTION TO PEDIATRIC PALLIATIVE CARE

***Dighe M**
****Muckaden MA**
*****Vora T**

Abstract: *Palliative care for children is a holistic approach aimed at improving the quality of life of children and families with a life limiting illness. This article describes the important principles of palliative care practice and the range of conditions where pediatric palliative care is applicable.*

Keywords: *Palliative care, Pediatric, Dying children*

The word “Palliative” is derived from the Latin word “Pallium” which is translated to mean “to cloak”. Palliative care is an approach that aims to “relieve suffering” caused by a disease process without aiming to cure the disease.

Palliative care for children and their families is defined by the World Health Organization¹ as follows:

- Palliative care for children is the active total care of the child’s body, mind and spirit and also involves giving support to the family.
- It begins when illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease.
- Health providers must evaluate and alleviate a child’s physical, psychological and social distress.
- Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited.

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Department of palliative care

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Tata Memorial Centre, Mumbai.

- It can be provided in tertiary care facilities, in community health centres and even in children’s homes.

The basic principles of palliative care are embodied in this definition and these are:

- Holistic care
- Care across the continuum of the disease process
- Rests upon a multi disciplinary team approach
- Applicable across varied settings

(A) Holistic care

Dame Cicely Saunders is the founder of the modern hospice movement which started in the UK in the 1960s. Dame Saunders was the first to conceptualize ‘total pain’, describing it as ‘the division of a whole experience into physical, emotional, social and spiritual components’². This concept can well be applied to all the aspects of palliative care practice.

For example, an eight year old child with an advanced osteosarcoma may have pain due to a large tumor. However his experience of “pain” or “suffering” may be understood better in terms of the child’s perception of his illness, his fears, aspirations, hopes, experiences of family members in caring and spiritual issues that may concern the child.

The psychological dimensions of suffering manifest as fear, feeling of sadness, guilt, frustration, depression, feeling a burden or feeling lonely. Besides children are seldom given an opportunity to voice their concerns and therefore to receive support.

Most chronic illnesses are associated with an alteration in the social functioning of the child. Some of these may be inability to attend school, play with peers, being viewed as “someone different”.

Spiritual issues are related to concerns about death, dying, relation with God and with loved ones. Often even young children may have spiritual concerns - for example children as young as five years may question about death and what happens to a person after death. Older children

with abstract cognitive abilities may have concerns quite similar to adults. It is important to remember that children will seldom speak of their fears and concerns unless they are actively given an opportunity to do so and the skill of a palliative care professional lies in being able to provide a safe and comfortable environment for the child to talk about all these issues which affect his experience of the illness.

The physical, psychosocial and spiritual dimensions of pain are closely interrelated. For example, depression, guilt and isolation worsen physical pain and vice versa. Similarly spiritual suffering will aggravate physical pain. The “holistic” approach to caring for children with life limiting illnesses is based on the importance of realizing the interconnections among various components and that each one of these must be appropriately addressed so as to provide optimal relief from symptoms.

(B) Care across the continuum of the disease process

In the past, palliative care was typically introduced to patients and families after all curative options were exhausted; thereby depriving patients of opportunities for appropriate symptom control, counseling, support and discussing palliative care options. In order to overcome this gap in care, the American Academy of Pediatrics recommended an “Integrated model of palliative and curative treatment”. In this approach, components of palliative care are offered at diagnosis and continued throughout the course of illness, whether the outcome ends in cure or death.³

Early interaction with a palliative care team may well enhance the quality of life of patients and the families by a)improving control of pain and other symptoms, b)better communication of information about the available treatment options (both curative and palliative) and c)provision of support in an individualized and empathic manner. If a point is reached in the disease trajectory when cure is no longer possible and only palliative care is to be provided, the transition becomes easier for patients and families if they are already familiar with the palliative care team through previous interactions.

An important component of palliative care is skilled care through the terminal phase and bereavement support. The Institute of Medicine defines a good death as “one that is free from avoidable suffering for patients, families and caregivers in general accordance with the patients’ and families’ wishes”⁴ In the pediatric setting, application of this definition may be challenged by certain issues such as a) the child’s ability to report suffering,

b) autonomy and decision making ability of young children and c) difficulty in predicting the time of death.

The death of child in particular, may lead to significant stress for (professional) caregivers, leading them to withdraw from dying child and his family.⁵ It is known that families are affected deeply by the attitudes of physicians and nurses in caring for their loved one in his final stages.⁵

Drawing upon recommendations that have been made for the elderly⁶, the basic principles to improve the care for dying children are given in the Table I.

Table I. Principles of a good death

- | |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <ul style="list-style-type: none"> • Being aware of dying (the family and the child wherever appropriate) • Being able to retain control of what happens (The child and the family) • Being afforded dignity and privacy • Having relief from pain and other symptoms • Having a choice about the place of care (at home, hospital or hospice) • Having access to information and expertise wherever necessary (including specialist palliative care, spiritual care, etc.) • Having control over who is present and who shares the end • Having time to say goodbye • Being able to die when it is time to go, and not to have life prolonged pointlessly |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|

Hence palliative care teams can help to improve the care received by dying children by a) becoming directly involved in the care wherever necessary, b) educating all health care professionals caring for children in the principles and c) practice of palliative care and being involved in broader advocacy for palliative care.

The intensive care setting: Intuitively it seems that palliative and intensive care are mutually exclusive. While the former focuses on comfort at the end of life, the latter focuses on intensive treatments to prolong life. However intensive care units provide care for morbidly ill patients and prognosis may be uncertain in some cases. Palliative care has the potential to improve care for patients by providing psychosocial support, pain control and support for stressed family members.

(C) Multi disciplinary approach

Palliative care aims to improve the quality of life of patients by addressing not only the physical but also psychological, social, spiritual aspects besides assisting in practical issues about caring for the patient.

Referring back to the example of the eight year old with osteosarcoma, the care of this child can be optimized with inputs of many different members of the pediatric palliative care team. Palliative amputation may be carried out to reduce the problems related to the child's painful useless limb. The child would receive support from the psychologist, nurse and the physician to receive information about the surgery in an age- appropriate and non frightening manner. The rehabilitation therapist can help this child in relearning to walk with the help of a prosthetic limb. This is a simplified framework of palliative care to improve the quality of life of the child.

Members of the pediatric palliative care team

The International Association of Hospice and Palliative Care⁷ recommends the coordinated involvement of the following professionals to deliver effective palliative care for adults-medical staff, nursing staff, physiotherapists, occupational therapist, dietician, psychologist, chaplain, family members and the patient. The patient is an important member of the team- who must receive information in an age appropriate manner and be encouraged to be involved in decision making about her care. The family members are also important members of the care giving team as they are frequently involved in physically caring for the child. The care plan must be developed with due consideration of the child's and the family's preferences thus making them important team members.

Unlike adults, children are helped by the use of non verbal communication methods. They also use modalities like distraction and play effectively in reducing their pain. In order to make the best use of these therapeutic modalities, the pediatric palliative care team must include play therapists, music therapists and child life specialists wherever they are available.

(D) Applicable across varied settings

Palliative care for children can be provided at the child's home, in the hospital or in the hospice. Most adults with life limiting illnesses prefer to receive care at home⁸, however the choice of the place of care may change as death approaches⁹. In some situations, the place of care may be dictated by practical issues rather than by preference. Often in India, patients with complex conditions

come to cities to receive treatment but as the disease advances, the families prefer to go home where they have the support of an extended family. The advantages of home based pediatric palliative care are - the patient and family continue to remain in the familiar and cherished environment of their home, support from family members is more accessible, parents and siblings can continue their work, schooling or other activities to some extent. The problems with home based palliative care are - lack of access to medical facilities in case of a medical emergency, overly reliant on the family to care for the patient, caregiver stress, poor living conditions.

Pediatric palliative care can also be provided by a specialist palliative care team or by appropriately trained hospital staff when the child is admitted for inpatient care. The advantage of the hospital setting with a palliative care unit is that multi disciplinary care is close at hand ensuring quick response to medical events and better continuity of care. This is particularly important when families have formed good relationships with the care team and value their presence and support. However hospital based care means significant disruption in the family's routine and usually increased cost of caring.

In the medieval period, the hospice was a "place of rest" for weary travelers. The modern hospice is a place of care for patients with life limiting illnesses where they may be admitted for respite care, relief of symptoms or for end of life care. The hospice has the advantages of the hospital with the comfort of a home. However with a limited number of hospices in the country, access to hospice based care is limited for many patients.

The ideal model of pediatric palliative care must incorporate all the above three components of care with seamless transfers across all three settings.

The scope of pediatric palliative care

Pediatric palliative care should be offered to all children (and families) who have a life limiting or life threatening condition. The Association for Children's Palliative Care (ACT) organization, UK, delineates four broad groups of life-threatening and life-limiting conditions.¹⁰ These categories are given below.

Category 1. Life-threatening conditions for which curative treatment may be feasible but can fail: Where access to palliative care services may be necessary when treatment fails or during an acute crisis, irrespective of the duration of that threat to life. On reaching long term remission or following successful curative treatment there is no longer a need for palliative care services.

Examples: Cancer, irreversible organ failures of heart, liver, kidney.

Category 2. Conditions where premature death is inevitable: Where there may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities.

Examples: Cystic fibrosis, Duchenne muscular dystrophy.

Category 3. Progressive conditions without curative treatment options: Where treatment is exclusively palliative and may commonly extend over many years.

Examples: Batten disease, mucopolysaccharidosis.

Category 4. Irreversible but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death.

Examples: Severe cerebral palsy, multiple disabilities such as following brain or spinal cord injury, complex health care needs and a high risk of an unpredictable life-threatening event or episode.

Preliminary studies from the UK have demonstrated that these categories are suitable tools to guide pediatric palliative care service development.¹¹ A comprehensive list of diagnoses which qualify for pediatric palliative care has been developed by experts based on the International Classification of Diseases 10. (Personal communication with Dr. Richard Hain, 2010). This list includes more than 400 such conditions.

The scope of palliative care for children is broad and it includes a wide range of varied pediatric disorders. The categories outlined above can serve as a useful guide to estimating the need for pediatric palliative care provision in a given setting, for policy makers to develop palliative care programs for children and for advocacy for children's palliative care.

Conclusion

Pediatric palliative care is applicable across different settings of care provision and a broad range of disease conditions. This multi disciplinary approach to caring will potentially improve the quality of life, of not only patients but also of families.

Ideally, all places caring for children with life limiting illnesses must have a pediatric palliative care team. However till such a time that this happens, basic palliative

care can be provided by pediatricians for their patients along with disease directed treatments. Inputs can be taken from specialist palliative care teams for caring for complex cases.

Points to Remember

- *Palliative care is an approach that aims to relieve suffering caused by a disease process without aiming to cure the disease.*
- *Early interaction with a palliative care team may well enhance the quality of life of patients and the families.*
- *In palliative care a multidisciplinary approach is a must.*

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SUPPORTIVE AND PALLIATIVE CARE

AN OVERVIEW AND RELEVANCE OF PEDIATRIC PALLIATIVE CARE IN INDIA

***Muckaden MA**

Abstract: *Pediatric palliative care is a relatively new field of speciality carved out between adult palliative care and pediatrics. It provides a holistic approach to care for children with life limiting illnesses. There are conditions defined which need such services which require team work. In India broadly these are cancer, HIV infections, thalassemia, neurological and other conditions. Palliative care aims at improving 'quality of life'. Past, present and future are depicted. Children's palliative care project for Maharashtra and it's implications are discussed.*

Keywords: *Children's palliative care, Holistic approach, Quality of life, Key role of professionals, Chronic illness.*

The goal of palliative care is to improve quality of life for people affected by life-threatening illness. The WHO¹ definition of palliative care is - 'An approach that improves the quality of life of patients and their families facing the problems associated with life-threatening illness, through prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual'.¹ The focus here is on improving quality of life when the disease becomes life-limiting. This necessarily means introduction of palliative care early in the trajectory of illness, so as to provide a smooth transition from active treatment to measures focused on quality of life (Fig.1).

Elements of palliative care

Palliative care is thus a patient and family-centered approach and focuses on the needs of the individual. Some key areas and their management are shown in Table.1.

Table I. Palliative care - Key areas and management

Physical	<ul style="list-style-type: none"> • Impeccable clinical assessment (including minimal investigations where appropriate) • Prevention and management of symptoms • Clinical treatment where appropriate • Appropriate referral to other clinics/hospitals if expertise is not available locally
Psychological and spiritual	<ul style="list-style-type: none"> • Assessment and management of psychological and emotional needs • Counseling - individual, family, bereavement support- Appropriate referral to other services as may be required • Where spiritual pain- referral to appropriate services (not necessarily religious)
Social	<ul style="list-style-type: none"> • Identification of financial needs • Developing social support networks • Support for the whole family especially where the traditional roles are altered • Appropriate referral to other services eg. NGOs
Legal	<ul style="list-style-type: none"> • Identification of legal requirements • Referral to appropriate legal services • Documentation of advance directives and patient preferences for care

(adapted from American Academy of Pediatrics, Committee on Bioethics and committee on Hospital Care²)

Palliative care for children

History

Most efforts to develop palliative care for children stemmed from efforts in the adult world. The very first unit

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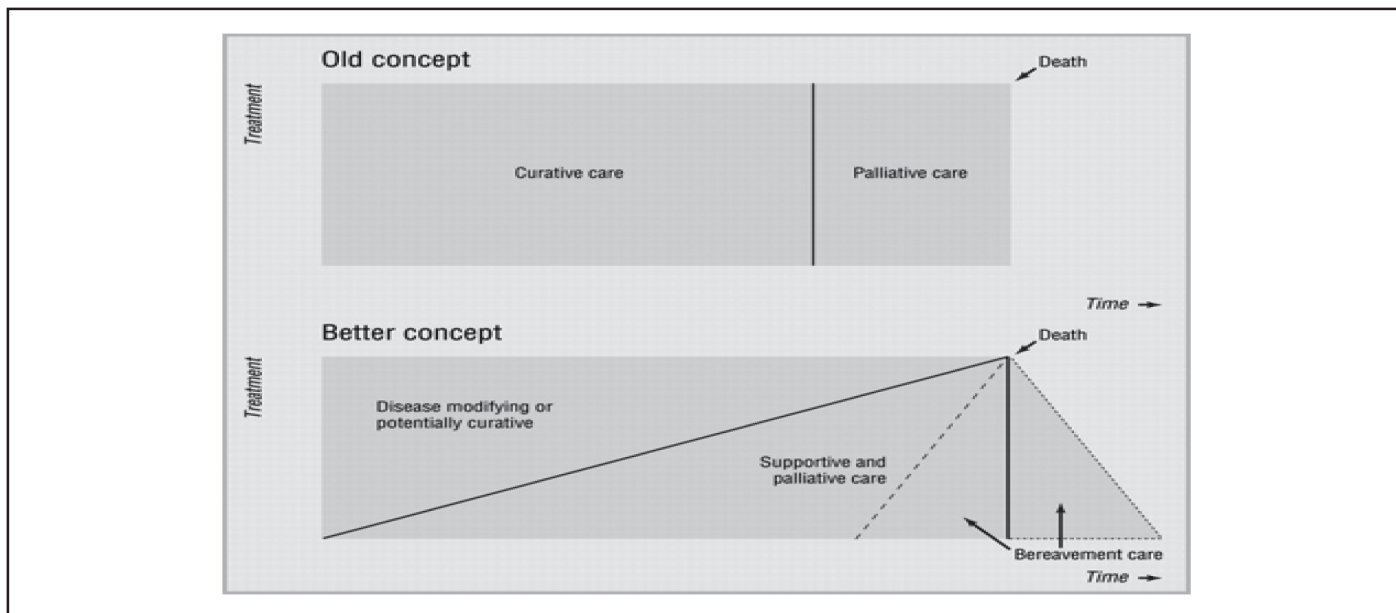


Fig.1. Disease trajectory and palliative care

Model for palliative care. (Reproduced from Murray SA, Kendall M, Boyd K, Sheikh A. Illness trajectories and palliative care. *Br Med J* 2005;330:1007–1011, with permission from BMJ Publishing Group Ltd.)

for pediatric palliative care was reported from St Mary's hospital in New York³ which was established in 1985 for children with cancer and AIDS. The first hospice movement was in the UK, with the development of the St Christopher's hospice in 1967 by Dame Cicely Saunders⁴ for adults and children with cancer. This model has been followed all over the Western subcontinent, for children with cancer and then with other life limiting conditions.^{5,6} At St Christopher's, within 1 year, a home care service was incorporated by Dr Mary Baines; utilizing community nursing and physicians to complete the circle of care at home. This proved even more relevant for the sick child in the midst of a large family. However, with the advent of nuclear families; hospices became relevant for 'respite care'; when the family had conflicting needs, necessitating alternate care for a specified period of time. The Helen House was the first such hospice for children set up in UK in the year 1982.⁷ Though even the founder had doubts about its utility initially there are now above 23 hospices in UK, with many more coming up in the UK, US, Europe and Australia. In the East, till date there are no reported dedicated hospices for children.

Conditions requiring palliative care

Children and young people with life-limiting or life-threatening conditions have very specific and unique palliative care needs, often different from those of adults. The above needs have to be tailored to the specific requirements of children and their families.

The Royal College of Pediatricians⁸ has divided causes of diseases in children needing palliative care under 4 main headings

1. Life threatening conditions for which curative treatment may be feasible but can fail eg. cancer, organ failure,
2. Conditions where premature death is inevitable but life may be prolonged eg. cystic fibrosis,
3. Progressive conditions without curative treatment eg. Batten disease,
4. Irreversible but non progressive conditions where severe disability can lead to premature death eg. Cerebral palsy.

Some of these conditions would cause distress to child and family for many years, even up to adulthood; thereby raising additional concerns whether to shift these children to the 'unfriendly' adult unit!

Global scenario

The International Children Palliative Care Network (ICPCN)⁹ estimates that worldwide twenty million children can benefit from palliative care, but access to such services remains an issue. Each year in the United States, approximately 500,000 children cope with life-threatening conditions and 53,000 children die from trauma, lethal congenital conditions, extreme pre-maturity, heritable disorders, or acquired illness. Less than one percent of children needing hospice services receive it in the United States. This is also true for most countries all over the world. The Association of Children's Hospices UK estimate

3,000 children using hospice services every year, the demand rising to 5,000 in the next 5 years. With the global surge of AIDS/HIV affecting populations, child mortality figures have increased drastically in developing countries due to mother-to-child transmission (MTCT). According to the UNAIDS/WHO Epidemic Update for 2005, out of the 3.1 million people who died from AIDS, over half a million were children under 15 years of age. It is estimated at the end of 2005, 2.3 million children in the world were living with HIV and 700,000 were newly infected. It is also a medical fact that HIV progresses to AIDS much more rapidly in children than adults, increasing their risk of dying in their very first years due to immuno-suppression. With most children having no access to treatment for HIV/AIDS, it is challenging to prevent infection and death of children in resource-limited settings and palliative care will have an increasing role in improving the quality of life of these children as well as the length of their life. It has been difficult to find statistics of children with the other conditions requiring palliative care, especially as reliable figures are not available from the developing world; the ICPCN statistics are probably the most reliable at the present moment in time.

Indian scenario

It is not easy to gather exact data regarding children needing palliative care in India. However, the few statistics available, estimate children with cancer needing palliative care as 55,000, with thalassemia 55,000; 55 506 children with HIV/AIDS in Mumbai (The 4th highest incidence of HIV/AIDS in India). Other common conditions requiring palliative care in India are severe mental retardation and cerebral palsy; neonatal severe diseases and death; severe cardiac and respiratory abnormalities, for which no estimates are available.

Needs of children (data gathered by International Children Palliative Care Network⁹)

- Children need appropriate pain and symptom management during the course of their illness. Studies have concluded that a vast majority of children with cancer need regular pain medication while in terminal care, whether it is administered orally or intravenously at home. Most pediatricians are reluctant to acknowledge that children suffer from pain and children being afraid of injections, also under report pain.
- Suffering from pain was more likely in children whose parents reported that the physician was not actively involved in providing end-of-life care.

- In some countries healthcare professionals never acknowledge death in children. The cultural denial of the fact that children actually die prohibits the development of children's palliative care services.
- The taboo around child death, without an open and honest approach in dealing with death and without adequate children's palliative care options, means that families are often forced to make inappropriate and ill-informed decisions to attempt aggressive curative treatments.
- Families should not have to choose between life-prolonging care and palliative care, when they can go hand-in-hand. There exists an assumption that palliative care should not be considered until all curative options are exhausted, when in fact palliative care should be seen as significantly improving a child's quality of life. It should be integrated with curative treatment, and throughout the course of the illness regardless of the child's outcome.
- Adult hospice and palliative care personnel may be able to provide expertise in end of life care for adults, but often have no pediatric expertise. Many of the conditions that are common in pediatrics are virtually unknown in adult palliative care. This decreases access for families and their children to health benefits such as home-based pain and symptom management, which should be individualized for specific family needs.
- Increased high-technology interventions do not always allow those in less well-developed countries to have access to basic health care services. Initiatives in developing countries should be community-orientated and sustainable.

Goals of children's palliative care

Some goals as estimated by ICPCN and other Children's Palliative Care organizations are

- To strengthen existing palliative care services for children.
- To develop palliative care services for children through integration into at least three existing projects and organizations in each country.
- To integrate palliative care policies for children into national palliative care associations.
- To improve access to HAART, palliative care drugs and pain medication for children.
- To advocate for the expansion of palliative care for children and its integration into government policies and national, regional and local health care systems.

- To educate health care practitioners in pediatric palliative care using a curriculum appropriate for developing countries.
- To develop materials on children's palliative care in local languages.
- To foster and protect the rights of children to life, education and sound health.

Mapping

As reported above, as the needs of children are increasingly being recognized; more and more dedicated pediatric palliative care services are coming up all over the world. The International Observatory on end of life care¹⁰ has conducted a survey for provision of palliative care services and findings are in Fig.2. Similar findings by ICPCN for children are depicted in Fig.3.

For India, the recognition that children and their families have very specific needs; has led to the development of specialized children's palliative care units. At present there are 4 such units all over India. They are at Can Kids Delhi (which was the 1st NGO to set up a dedicated support group for kids), MNJ Cancer Centre, Hyderabad, where the pediatric oncology unit and palliative care services work closely together to provide supportive care to children with cancer. At the Institute of Palliative Medicine, Kozhikode a dedicated unit has been recently established, along with a unit in the Dept of Pediatrics, Medical college, Kozhikode.

At the Tata Memorial Centre, Mumbai the 1st dedicated pediatric palliative care unit was set up in September 2002, to care for children and their families with advanced cancer. In Chennai, an NGO, Dean Foundation, has set up a Centre for Palliative Care at the Institute of Child Health and Hospital for Children, Egmore. There are also some new ones. At all adult palliative care units, children and their families are taken care of by trained professional teams.

Place of Care

Palliative care depends very much on background, culture and availability of resources. In developed countries, community services are very well developed and care for the child and family is undertaken by the community services in liaison with the specialist palliative care services, wherever necessary. In countries like India, the community services are very limited; this even more so in rural areas. However, experience at the Tata Memorial Centre has shown the over-riding preference for families to take care of their ailing child at home, even in the remote village. Here then, we need to utilize whatever resources that are already available. The existing health care systems in the country, both governmental and private need to be harnessed, especially as a private-public partnership. There is a role for doctors from alternative medicine, ANM nurses and the family members themselves, all of whom can be trained to care for the child. This approach can assist in the child remaining in familiar surroundings till the end.

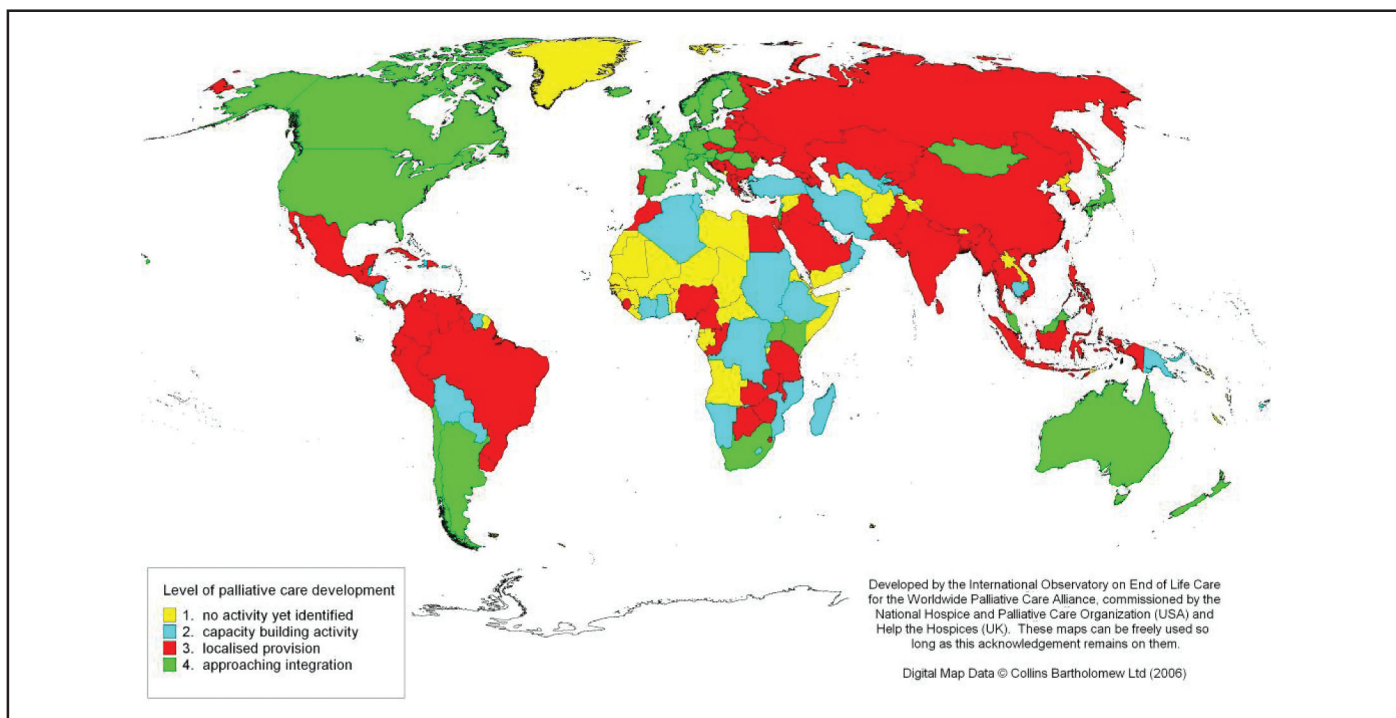


Fig.2. Data compiled by international observatory on end of life care 2006

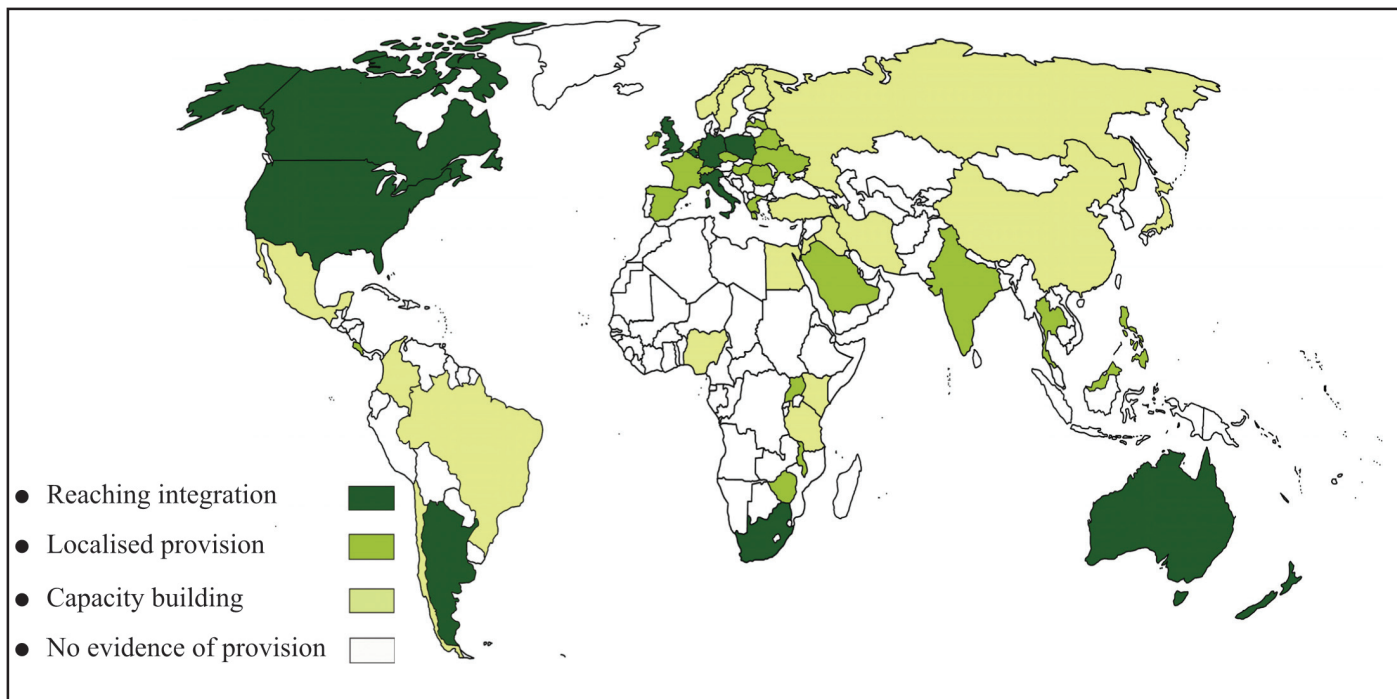


Fig.3. ICPCN map estimating levels of children's palliative care service provision worldwide

It is also important to focus on the fact that the ideal place of care can change as the situation evolves. The ideal place can thus shift from home to hospital and back to home again. The decision to change the place of care should be discussed at every stage with parents, physician, nurse and if possible the child itself.

Place of death is again unique for every culture and the same decision making process should be followed. In our experience, parents feel safe in the hospital setting as the surroundings are familiar. It is important that goals of care are meticulously reviewed as often as necessary as decision making for the family is very emotive at this stage. 'Quality of life' and 'death' are best achieved in a corner of the ward or side room with the family at the bedside with a minimum of intervention to keep the child as comfortable as possible. Parents will always remember the empathy of the staff.

Culture and Children Palliative Care (CPC)

Palliative care of children is also very culture specific. Needs of the family are intertwined with their individual religious beliefs. Though there is always a hope for 'cure', yet the acceptance of 'karma' as tradition allows parents some ability to accept poor prognosis when given with empathy. Supporting them all through the 'phases of grief' is the duty of all palliative care professionals.¹¹

The size and structure of families also dictate the role of the professional. Where families are mainly nuclear;

family members are few and there is much more reliance on professional carers. Here the availability of hospice care becomes an integral component. Where families are large; there are always available carers. Training of family members for medication and basic needs, allows children to be cared for in the midst of family even in the absence of community nursing. In Eastern cultures, neighbours and friends, also form part of the caring team for patients or siblings. Home care is thus somewhat culture specific.

Knowledge about 'end of life' care is also necessary for palliative care teams. The member may even be called upon to arrange the specific rituals for a family, in strange surroundings. In Western countries, a chaplain is always available; in India, religious leaders do not always visit hospitals. A home death would need the arrangement of the death certificate and other formalities. This is best arranged in anticipation, with a visit to the local GP. After the death, the team members play a large role in helping the family move on.

Future of palliative care of children

As more centers are being developed for CPC the specific needs of the community have to be kept in mind. Children needing the services are scattered; dedicated services are not always feasible. For developed countries; more children hospices are being developed. In the Eastern culture, a hospital based CPC is being developed with home care being one of the key components. There will thus always be role for the involvement of both the adult palliative

care units and the pediatricians to optimize resources to develop effective CPC in India.

Pilot project for children's palliative care in Maharashtra

In October 2010 the UK Department for International Development (DFID) approved a proposal to improve access to palliative care for children with HIV/AIDS in Malawi and the Maharashtra District of India, over a five year period. The Tata Memorial Hospital, with its expertise in Pediatric Palliative Care for children with cancer, is supporting the children's palliative care project with their mentorship. ICPCN, Department for International Development (DFID) and Help the Hospices are partners of the project.

The project is advocacy based to gain government approval for pediatric palliative care in India. This was envisaged with the development of 3 sites, to serve as models for further development in the State

1. Lokmanya Tilak Municipal General Hospital (L.T.M.G.H) Sion, Mumbai: The Regional Pediatric AIDS Centre at LTMG Hospital is a National Centre of Excellence for Paediatric HIV and has around 1785 registered children. The aim for this centre was to introduce palliative care for these families; to provide a holistic approach including psycho-social care for children with HIV. More than 40 children have been included in CPC and have been provided various types of interventions.

2. Jawar cottage rural hospital, Thane: This site was set up for training and awareness on CPC in the rural part of Maharashtra. The program is community based. ASHA workers of NRHM project act as a liaison for referral of patients from community to PHC's and cottage hospital. All the existing doctors, nurses and health care workers have been trained in palliative care. One doctor and social worker have been appointed only for this project. This is a good example of utilizing existing resources.

The implementation of the project is objective based including monitoring and evaluation with these main objectives:

- 1. Adoption of pediatric palliative care policy by the government** (Involvement of National and International Organizations and Health Ministry)
- 2. Training and awareness** to doctors (mainly pediatricians), nurses, social workers, health care professionals, volunteers and NGO's in knowledge, attitude and skill to deliver supportive care to children.

3. Enhance availability to pain relieving medication especially morphine

4. Empowerment of children and families for improving quality of life

Some achievements up to date are

1. The two sites in Sion and Jawar are functioning well and act as model sites for CPC
2. The pediatric palliative care policy for Maharashtra has been submitted to the State Govt. officials and is under scrutiny.
3. Many pediatricians and other health staff have already been trained in CPC, we look forward to this continuing steadily over the next 3 years.
4. With this project the NRHM had adopted palliative care and CPC at 1 site and we look forward to developing many more sites.
5. Morphine is made available at Jawar for those who need it within the existing Narcotic drugs and psychotropic substances (NDPS) rules.
6. Results of focus group meetings with affected children and their parents will be incorporated into fact sheets for Govt. and professionals.

The biggest achievement of this project will be the adoption of principles of children's palliative care by pediatricians in India for children with life limiting conditions.

For further information on the project visit the website at [www.palliativecare.in/ children's palliative care](http://www.palliativecare.in/childrens-palliative-care).

For any further information and training please contact the author at muckadenma@tmc.gov.in or childrens-palliativecare@gmail.com.

Points to Remember

- *Children with life-limiting or life-threatening conditions have very specific and unique palliative care needs, often different from those of the adults.*
- *The focus is on improving 'Quality of life' which requires introduction of palliative care early in the trajectory of illness.*
- *Children need appropriate pain and symptom management during the course of their illness. This includes not only physical care, but psychological, social and spiritual care for the child and family.*
- *This requires country specific education and training for all cadres of health care practitioners in paediatric palliative care*

- ***There is a need for the expansion of palliative care for children and its integration into government policies and national, regional and local health care systems.***

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CLIPPINGS

Reduced Risk of Pertussis Among Persons Ever Vaccinated With Whole Cell Pertussis Vaccine Compared to Recipients of Acellular Pertussis Vaccines in a Large US Cohort

Unexpected waning of immunity after pertussis vaccination is now well described. In this study we examined whether prior vaccination with whole-cell pertussis vaccine (wP) at any point provided superior protection contrasted with a solely acellular pertussis vaccine (aP) series. We utilized the coincidence of a large outbreak of pertussis with the termination of wP availability, providing populations of children who had been vaccinated with combinations of wP and aP.

Kaiser Permanente (KP) is an integrated healthcare system with complete electronic records and a centralized laboratory. Cases of laboratory-confirmed pertussis and vaccination data for members aged 8–20 years were retrieved.

Among 263 496 persons aged 8-20 years, 904 cases of pertussis were identified. In patients with a full history of vaccinations administered by KP, those with 5 total doses of only aP had an 8.57 relative risk (RR) of pertussis ($P < 0.0001$) contrasted to those with ≥ 1 wP dose. With 6 doses of aP, the RR of disease was 3.55 ($P < 0.0001$). When external vaccine records were included, the results were similar.

We found a markedly increased risk of disease associated with an entirely aP series. This risk was mitigated, but not eliminated, by the presence of a sixth dose of pertussis vaccine (Tdap). Receipt of one or more wP doses markedly augmented the durability of immunity from subsequent aP dose. It appears that a wholly acellular pertussis vaccine series is significantly less effective and durable than one that contains the traditional whole cell vaccine.

Maxwell A. Witt, Larry Arias, Paul H. Katz, Elizabeth T. Truong and David J. Witt. Clin Infect Dis. (2013) 56 (9): 1248-1254. doi: 10.1093/cid/cit046. First published online: March 13, 2013.

SUPPORTIVE AND PALLIATIVE CARE

ORGANISING A PEDIATRIC PALLIATIVE CARE UNIT

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Abstract: *The essentials of pediatric palliative care unit is the skilled and interdisciplinary attention to pain and other distressing symptoms; emotional, spiritual and practical support; assistance with complex medical decision-making; and coordination across the continuum of care settings. Though the principle of care and basic structure remains the same, organizing a pediatric palliative care in rural and urban setting differs a lot. It definitely depends on background, culture and availability of resources.*

Keywords: *Pediatric palliative care, Children, Multi disciplinary team, Place of care.*

“Regardless of their age, children suffer all the clinical, psychological, ethical and spiritual aspects of incurable illness and death. However, they are not small adults; they have specific developmental, psychological, social and clinical needs that must be addressed.” - Vittorio Ventafridda

In India, out of 75,000 children with recorded cancer every year, less than 20% survive. Many more probably die before a diagnosis is made. Furthermore, among the estimated 2.5 million people in India living with HIV/AIDS, 70,000 are children under the age of 15 years.¹ These children, as well as many others with life limiting diseases, need help and support to reduce their morbidity. It is estimated, however, that at present less than 1% of them have any access to palliative care.²

With the advancement in the medical research, the scenario of management of many life limiting conditions is changing. Still lot of issues are to be addressed in pediatric palliative care for which an ideal set up is envisaged.

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An ideal model for pediatric palliative care

According to the guidelines of WHO, pediatric palliative care starts when the illness is diagnosed.

According to American Academy of Pediatrics (AAP)³,

- Palliative care and respite programs need to be developed and widely available to provide intensive symptom management and promote the welfare of children living with life-threatening or terminal conditions.
- At diagnosis of a life-threatening or terminal condition, it is important to offer an integrated model of palliative care that continues throughout the course of illness, regardless of the outcome.
- All general and subspecialty pediatricians, family physicians, pain specialists and pediatric surgeons need to become familiar and comfortable with the provision of palliative care to children. Residency, fellowship training and continuing education programs should include topics such as palliative medicine, communication skills, grief and loss, managing prognostic uncertainty, decisions to forego life-sustaining medical treatment, spiritual dimensions of life and illness and alternative medicine.^{4,5,6}
- An increase in support for research into effective pediatric palliative care programming, regulation and reimbursement, pain and symptom management and grief and bereavement counseling is necessary. The pharmaceutical industry must provide labeling information about symptom-relieving medications in the pediatric population and provide suitable formulations for use by children.
- The practice of physician-assisted suicide or euthanasia for children should not be supported.

The UK Government Health Departments has published strategies to develop the palliative care services received by children, young people and their families. UK children's palliative care charity, ACT (Association for Children's Palliative Care), has prepared the following quality markers for children's palliative care services.⁷

The services should have

- An action plan for the delivery of high quality palliative care, which is reviewed for impact and progress.
- It should encompass children, young people (including those in transition to adult services) and their families.
- Implement effective mechanisms to identify children and young people with life-limiting or life-threatening conditions at any point from the time of diagnosis or recognition right through to the approach of their end of life phase.
- Ensure that the child or young person and their family's preferences and choices are documented and communicated to appropriate professionals and that the needs of family members, including siblings are appropriately assessed and recorded through a carer's assessment.
- Have mechanisms in place to ensure that care for children and young people is co-ordinated across organisational boundaries 24/7.
- Ensure that all children and young people who are approaching end-of-life are offered a care plan.
- Have sustainable essential services available 24/7 to all children and young people who are approaching end of life. These services should also be capable of offering home-based care at end of life, if this is what the family chooses.
- Be aware of children's palliative care and end of life care training opportunities and enable relevant workers to access or attend programs appropriate to their needs.
- Adopt a standardised approach to care for life-limited children and young people throughout their whole care journey.
- Ensure mechanisms are in place to provide effective feedback and meaningful evaluation from parents and carers.
- Monitor the quality and outputs of children's palliative care including end of life care, and submit relevant information for local and national audits.

The aims of pediatric palliative care are to

- Provide family-centered care in both community and hospital settings for children with life-limiting illness
- Offer support and advice for the child, family and staff
- Provide education for the child, parents/carers, family, schools and professionals in the community, hospital and voluntary sectors

- Co-ordinate care between home, clinic, hospital and respite/residential services, liaising with fellow professionals as appropriate
- Provide specific advice on and treatment of pain and symptoms
- Assist with care of the child at home or in hospital in the terminal stages of their illness in collaboration with the family, the multidisciplinary team and primary care
- Provide bereavement support for family members and fellow professionals
- Provide an evidence-based resource centre for pediatric palliative care

Essential requirements of pediatric palliative care unit

Palliative care for children can be provided at the child's home, in the hospital or in the hospice.

1. Multidisciplinary team

A pediatric palliative care set up should be hospital or clinic based where families of children with life limiting conditions have been coming for the services. The children's palliative care team should ensure a high standard of care and support for children with life-limiting conditions and their families. The team should work through primary care teams and with hospital staff to ensure consistency and continuity of care and to maintain awareness of good practice. The team should act as a resource for staff and healthcare workers from other institutes and voluntary organizations. The team should ideally consist of a pediatrician, clinical child psychologist, social worker/s, trained nurse/s. Other professionals are co-opted to the team as and when required, i.e. pain specialist, dietician, chaplain, play therapists, music therapists and child life specialists. Although progress has been made in medical management of many childhood conditions, this has not always been accompanied by a similar consideration for the emotional and psychological wellbeing of the child and family. This lack of attention often results in unnecessary pain, anxiety and suffering. Hence a set up should comprise of the multidisciplinary specialists.

2. Training and education

Health care workers should be trained to identify child's need for palliative care, to assess emotional and spiritual needs of the child and the family to facilitate advance care planning, to assess and manage child's pain and other symptoms, bereavement support to child's family

and to recognize the need to refer the child to a specialized care.⁸ The interdisciplinary team should be equipped with knowledge and skill in ethical, legal and regulatory aspects of medical decision-making.

3. Dedicated staff

The staff should be empathetic and dedicated for the welfare of the child and the family. All the healthcare workers should be ready to spend time for patients. They should be comfortable with the care for dying children. In consultation with the child's parent or guardian, the plan of care incorporates respect for the terminally ill child's preferences concerning testing, monitoring and treatment. The needs of families must be attended to both during the illness and after the child's death.

In addition to alleviating pain and other physical symptoms, physicians must provide access to therapies that are likely to improve the child's quality of life.⁹ Such therapies may include education, grief and family counseling, peer support, music therapy, child life intervention or spiritual support for both the patient and siblings and appropriate respite care. Respite care, the provision of care to an ill child by qualified caregivers other than family members, allows the family time to rest and renew, whether for hours or days, on a schedule, or intermittently as needed.

4. Availability of drugs for symptom management

The pediatric palliative care unit should store all the essential drugs necessary for the pain and symptom management. The unit must have license to dispense and store morphine and other required drugs. A proper documentation of dispensing has to be kept by the concerned responsible pharmacist/nurse.

5. Home care team

Home care facility eliminates the need for care in a hospital or care institution. This can be helpful to the children living in rural or remote area and for those who cannot access hospital care due to financial restraints or caregivers' inability to accompany them due to fear of losing daily wages. It also provides preventive medicine and assists in relieving stress of caregivers. Home care service encourages a high degree of participation by the children receiving care since it is their familiar environment. Its support from family members is more accessible, parents and siblings can continue their work, schooling or other activities to some extent.

6. Networking

A local network of specialist support on site and a

24/7 on-call service are necessary in order to facilitate the implementation of basic palliative care. A setting up of networks for palliative care for children and adolescents and providing continuous care throughout a wide range of settings is an essential thing. Networking with voluntary organizations working for sick and needy children, support groups, spiritual experts, orphanages and hospices will help the children in getting various services and facilities to improve their quality of life. Networking and training of community care workers i.e. ANMs and ASHA workers can help in identifying needs of children with life limiting conditions.

7. Transport facilities

Ideally the unit should have a transport facility for home care team and to take the sick child to any specialized care services.

8. Proper documentation

Efficient, comprehensive documentation is a vital element of all healthcare provision. It not only provides a record of care, but should reflect the quality of the care, enable continuity of care between practitioners and reinforce care standards.¹⁰ Case record forms should include all the details i.e. demographic data, social assessment, psychological and behavioral assessment, clinical, symptom, nursing and nutritional assessment and follow up sheets. A proper documentation should be kept about dispensing of essential drugs and other services provided to the children.

9. Facilities to deal with emotional stress of the staff

Health care workers must be supported by the palliative care team, their colleagues, and institutions in dealing with the child's dying process and death. Institutional support may include paid funeral leave, routine counseling with a trained peer or psychologist, and regularly scheduled remembrance ceremonies or other interventions such as inviting bereaved families to return and celebrate with staff the deceased child's life.¹¹

10. Improved professional and social support for pediatric palliative care

Regulatory, financial and educational barriers often bar families from access to pediatric palliative care services. Professional and public education may foster awareness of the need for, and value of, pediatric palliative care and lead to efforts to remove bureaucratic and economic obstacles to its availability.¹¹

11. Continued improvement of pediatric palliative care through research and education

Clinical research concerning the effectiveness and benefits of pediatric palliative care interventions and models of service provision should be promoted. In addition, information about pediatric palliative care that is already available must be effectively disseminated and incorporated into education and practice.¹²

Place of care

Though the principle of care and basic structure remains the same, organizing a pediatric palliative care in rural and urban setting differs a lot. It definitely depends on background, culture and availability of resources. Developing a pediatric palliative care services in urban areas is more easier since the system is already developed. Rural areas typically are characterized by the lack of well-organized services, with primary care professionals and community nurses, having to undertake the care. The challenges involved in organizing pediatric palliative care in rural areas compared with urban areas relate to differences in environment, infrastructure and resources.^{13,14} The population is thinly distributed over large areas, with few specialists.¹³ Consequently, primary care professionals and nurses, play crucial roles in both organizing and delivering palliative care services.

In developed countries, community services are very well developed and care for the child and family is undertaken by the community services in liaison with the specialist palliative care services, wherever necessary. In counties like India, the community services are very limited; even more so in rural areas. In such scenario, the available resources should be utilized. Due to the varied geographical conditions, it is difficult for patients as well as the health care workers to access or provide the health care facilities.

In rural settings, community care workers play an important role in this settings. In India they are called ASHA (Accredited Social Health Activist). One of the key components of the National Rural Health Mission in India is to provide every village in the country with a trained female community health activist. They are selected from the village itself and accountable to it, and are trained to work as an interface between the community and the public health system. They do the surveillance of the houses allotted to them. So they are well aware of the physical, psychological and social issues of the family. This can be helpful to identify the children who may need palliative care. They can link with nearest functional health facility for referral services and can identify transport for referral of

cases from village to facility. There is a role for doctors from alternative medicine, ANM nurses and the family; all of whom can be trained to care for the child. This approach can assist in the child staying in familiar surroundings till the end.

Points to Remember

- *The goal should be to assist with the care needs of children and their families to achieve the best possible quality of life in accordance with their values, preferences and beliefs.*
- *Palliative care needs in children are varied and according to the respective diagnosis and illness trajectory different approaches and measures are indicated.*
- *As medical and technological advances reduce childhood mortality and improve survival for children with life-limiting conditions, there is a need to integrate effective and efficient child-specific palliative care into national health and social service policy.*
- *It is very important to integrate specialized services like palliative care into the existing health care system for maximum utilization with minimum of cost; yet providing specialty services.*
- *It is also important to focus on the fact that the ideal place of care can change as the situation evolves. The ideal place can thus shift from home to hospital and back to home again. The decision to change the place of care should be discussed at every stage with parents, physician, nurse and if possible the child itself.*
- *An integrated model of palliative care “in which the components of palliative care are offered at diagnosis and continued throughout the course of illness, whether the outcome ends in cure or death” has to be adopted.*

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CLIPPINGS

Early cord clamping versus delayed cord clamping or cord milking for preterm babies

Optimal timing for clamping the umbilical cord at preterm birth is unclear. Early clamping allows for immediate transfer of the infant to the neonatologist. Delaying clamping allows blood flow between the placenta, the umbilical cord and the baby to continue. The blood which transfers to the baby between birth and cord clamping is called placental transfusion. Placental transfusion may improve circulating volume at birth, which may in turn improve outcome for preterm infants.

Objective of the analysis was to assess the short- and long-term effects of early rather than delaying clamping or milking of the umbilical cord for infants born at less than 37 completed weeks' gestation, and their mothers. Providing additional placental blood to the preterm baby by either delaying cord clamping for 30 to 120 seconds, rather than early clamping, seems to be associated with less need for transfusion, better circulatory stability, less intraventricular haemorrhage (all grades) and lower risk for necrotising enterocolitis. However, there were insufficient data for reliable conclusions about the comparative effects on any of the primary outcomes for this review.

Rabe H, Diaz-Rossello JL, Duley L, Dowswell T. Effect of timing of umbilical cord clamping and other strategies to influence placental transfusion at preterm birth on maternal and infant outcomes. Cochrane Database of Systematic Reviews 2012, Issue 8. Art. No.: CD003248. DOI: 10.1002/14651858.CD003248.pub3. Published Online: August 15, 2012.

NEWS AND NOTES

42nd Annual Conference of Indian Academy of Pediatrics – Kerala State

Dates: 4th, 5th & 6th October, 2013

Contact

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SUPPORTIVE AND PALLIATIVE CARE

PRENATAL PERSPECTIVES IN PALLIATIVE CARE

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Abstract: *Palliative care for neonates is “an entire milieu of care to prevent and relieve infant suffering and improve the conditions of the infant’s living and dying.” In pregnancies with a malformed fetus, the parents and the live born will benefit from early involvement of palliative care professionals. In this article we attempt to discuss the important issues related to the care of a mother carrying a malformed fetus.*

Keywords: *Prenatal conditions, Perinatal palliative care*

Congenital anomalies refer to structural defects (congenital malformations, deformations, disruptions and dysplasias), chromosomal abnormalities, inborn errors of metabolism and hereditary disease present at birth.¹ They contribute to a significant proportion of childhood deaths in the developed countries. With better access to ultrasound and other techniques, the prenatal detection of congenital anomalies has increased² resulting in challenging care needs of parents and of the live born fetus with a life threatening anomaly. In this chapter, we attempt to describe the role for palliative care in cases of prenatally detected anomalies.

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According to Catlin and Carter, palliative care for neonates is “an entire milieu of care to prevent and relieve infant suffering and improve the conditions of the infant’s living and dying.”³

It is a “team-based” approach also aimed at relieving the psychosocial, emotional and spiritual suffering of the family. In cases where the delivery of an extremely premature or malformed baby is anticipated, the palliative care approach may well be applied prior to the birth. Along with the primary team of obstetricians and neonatologists, the palliative care team can assist the parents or family in making decisions about care (eg. resuscitation) for the newborn, place of care and provide psychological support. Post delivery, the palliative care team can provide comfort care to the baby, after death care and bereavement support to the family whichever is needed.

Epidemiological considerations

In 1999, congenital anomalies were responsible for approximately 12% of all childhood deaths in the USA.⁴ In Europe, major congenital malformations are reported in at least 2% of all fetuses and infants.⁵

In the period 1995–1999, data were obtained from 17 European population-based registries of congenital malformations (EUROCAT). In this study, 11 severe structural malformations that can be identified by ultrasound were included and the outcomes were noted. 68% anomalies were detected before 24 weeks of gestation. 43% of 4366 cases diagnosed with the 11 severe structural malformations across 17 registries resulted in termination of pregnancy while 53% resulted in live born babies with a median gestational age of 37 weeks for prenatally detected cases.⁶

Data from EUROCAT study have definite implications for provision of palliative care for babies detected to have congenital anomalies in the antenatal or the post natal period and these are discussed in the subsequent paragraphs.

Psychological issues in pregnancy

Pregnancy leads to several profound physiological and emotional changes in the mother. Women experience ambivalence, anxiety, a heightened sense of vulnerability and concerns for the safety of the unborn child.⁷

Mothers experience intense grief reactions following either giving birth to a child with a life-limiting condition or termination of pregnancy for fetal anomaly.⁸ When a congenital malformation is detected prenatally, the mother experiences acute psychological distress.⁹

Fathers who have experienced a perinatal loss are known to have intense feelings of loneliness, isolation and pain but they are less likely to be expressive about their feelings.¹⁰ The bereavement for fathers may be compounded by the expectation that they manifest strength and support which is often in conflict with his own sense of loss.¹¹

Besides the parents, the siblings and extended family members such as the grandparents also need support.

Therefore it is evident that parents and families of babies who have congenital malformations have significant emotional needs and palliative care teams can play an important role in providing such support.

Detection of congenital anomalies

Ultrasound and other prenatal techniques have increased the rate of detection of severe anomalies. Anencephaly, gastroschisis are some of the most commonly detected anomalies (approx. 90%) whereas transposition of great vessels and limb reduction defects have detection rates of approximately 30%.⁶ In the EUROCAT study which assessed the incidence of only 11 severe malformations, 68% were detected before 24 weeks.⁶

Once a mother is detected to have a fetus with a potentially life threatening condition, the implications for care are complex. Some of the significant issues are discussed below.

Breaking bad news

Any news that drastically and negatively changes a person’s perspective of his life are bad news. Breaking bad news to parents when the fetus is detected to have an anomaly needs skill and compassion.

Health care professionals must realize that they cannot make “bad news” into a “good news”; but they can convey the news in a sensitive manner-which families will value for a life time. Some of the factors that facilitate or hinder the practice of delivering bad news are given in Table I.

Bad news may be broken to the parents by the primary obstetrician while the palliative care team and the neonatologists are available to provide any additional support that may be required.

Prognostication

Nearly 6% of babies born with a congenital anomaly die in the first week of life; however the prognosis is quite disparate for different types of anomalies. The median survival time for all ascertained cases who died was 11 days. The shortest median survival time for those who died was in cases with urogenital tract and kidney anomalies, musculoskeletal and connective tissue anomalies and respiratory system anomalies.⁴

While the prognosis for some conditions like anencephaly and renal agenesis are clear, several conditions do not have a clear cut straight forward prognosis.

Counseling parents in the antenatal period must take the prognosis for survival into account. In situations where it is difficult to ascertain the survival outcome, it is advisable to acknowledge the uncertainty while remaining focused on the quality of the baby’s life.

Table I. Factors facilitate and hinder breaking bad news

Factors that facilitate breaking bad news	Factors that hinder breaking bad news
<ul style="list-style-type: none"> • Privacy, making time • Open ended questions • Finding out what the family knows • Using supportive non verbal cues • Not minimizing family’s concerns • Providing emotional support • Empathy • Assuring family of continuation of care and follow up 	<ul style="list-style-type: none"> • Lack of these • Closed questions • Assuming • Not allowing the family to talk or ask questions • “Side stepping” difficult or emotional issues • Giving false hope • Conveying that we are “not available” • Saying “there is nothing I can do for you.”

Termination of pregnancy

Termination of the pregnancy may be offered to couples after the detection of a severe congenital anomaly. However there are no standard guidelines and the procedure is governed by laws which are different for different countries. Physicians must provide adequate information and support to parents so as to help them make appropriate decisions.

Termination of pregnancy for a fetal anomaly rate (TOPFA) across nineteen registries of congenital anomaly in 12 European countries between 2000 and 2005 was 3.4 per 1000 births and 0.25 beyond 25 weeks.¹² The median time interval from diagnosis to termination was 2 weeks probably reflecting the time period needed to allow referral to specialists and prenatal counselors, to wait for karyotype results, to allow parents to reach an informed decision and to obtain permission for termination from a committee or more than one doctor. In four anomalies viz diaphragmatic hernia, omphalocele, arthrogryposis multiplex and Turner's syndrome, the time was longer probably because of the time required to assess secondary problems arising due to the anomaly.¹²

In a study from the US where a perinatal hospice was available, it was found that the structured program providing ongoing, comprehensive, multidisciplinary, supportive perinatal care offered a tangible and safe alternative to early elective pregnancy termination for patients carrying a fetus with a lethal congenital condition.¹³

Care planning and neonatal resuscitation

Parents must be involved in discussions about the starting of resuscitation when the need is anticipated at birth. The American Academy of Pediatrics¹⁴ makes the following recommendations for withholding or withdrawing neonatal resuscitation

- When gestation, birth weight, or congenital anomalies are associated with almost certain early death and when unacceptably high morbidity is likely among the rare survivors, resuscitation is not indicated. Examples include extreme prematurity
- In conditions associated with uncertain prognosis in which survival is borderline, the morbidity rate is relatively high and the anticipated burden to the child is high, parental desires concerning initiation of resuscitation should be supported
- In a newly born baby with no detectable heart beat, it is appropriate to consider stopping resuscitation if the heart rate remains undetectable for 10 minutes

Parents must receive adequate information about the prognosis of the resuscitation efforts and must receive appropriate emotional support to make decisions.

Place of care

Parents and families must be given opportunities before the baby's birth to decide about the place of care for the baby. The home has advantages of being the cherished environment where the family can spend precious time with the baby. However these families must have access to medical care whenever required.

If the family chooses to stay in hospital, their decision must be respected. However unnecessary investigations and intrusive therapies must be avoided so as to maximize the baby's comfort and avoid raising false hopes for the family. Parents, siblings and other family members must have the opportunities to visit the baby and have adequate privacy.

The parents, siblings and other family members must have opportunities to hold the baby, spend time with the infant and be able to collect mementos like footprints. The time spent with the babies and mementos may help families in their bereavement.

Spiritual needs

Families of dying children may have significant spiritual distress. Some of the issues may be as follows- Why me?, This is my destiny/karma, Questioning relationships with God, etc.

In the West, formal spiritual support may be offered by spiritual counselors or chaplains. This cadre of workers is currently available in India and they have an important role in the palliative care team.

Comfort care for the baby

The principles of care are similar to those for an older child. Simple comfort measures, such as positioning the baby with suitable boundaries, gentle rocking and swaddling, can be very effective in relieving discomfort.

The principles for the management of dying infants are adapted from the basic symptom control in pediatric palliative care. The rainbows children's hospice guidelines.¹⁵

Feeding: The aim of feeding the baby is to reduce distress from hunger and to ensure some hydration. Oral feeding must be encouraged as far as possible.

Pain: All babies who need palliative care must be monitored for pain. Some signs that indicate pain are persistent crying,

furrowing or bulging of the brow, furrowing of the nasolabial folds and tight squeezing of the eyes.

The recommended drugs for the management of pain are paracetamol or ibuprofen for mild pain and morphine for more severe pain. Drugs can be given orally or rectally.

Morphine: For acute pain, morphine can be given intravenously at starting doses of 10-20 µgm/kg/hour and is then titrated to response. Orally morphine must be given as a liquid formulation with appropriate dose for breakthrough pain. The starting dose is double the IV dose.

For babies who are dying other common symptoms are seizures, gastro-esophageal reflux, excessive secretions and irritability. These symptoms must be managed as for any other neonate. Sedatives may be required to help a baby to sleep; chloral hydrate orally or rectally is the preferred drug.

Siblings

The baby's siblings may often be forgotten. Siblings must receive information in an age appropriate manner. They must be encouraged to participate in caring for the baby in any way that they can. Feelings of guilt and other negative feelings must be dealt with by the health care team and opportunities for ventilation must be provided.

Bereavement support

Bereavement is a loss and the death of a child is one of the most tragic bereavements. Parents who lose a child suffer marked grief responses. Support in the bereavement period is aimed at helping the grieving family to cope with the loss and in identifying pathological grief.

The principles of bereavement support¹⁶ are given below.

1. Make contact and assess the bereaved parents
2. Provide assurance that they can survive their loss
3. Provide times to grieve
4. Facilitate the identification and expression of feelings, including anger, hostility, sadness, relief and guilt
5. Encourage verbalization of thoughts and recollections of the deceased child; do not be afraid to mention the deceased child's name
6. Interpret "normal" grieving behaviour and responses
7. Maintain a therapeutic and realistic perspective; do not rush to "fix" the pain

8. Allow for individual differences relating to gender, age, personality, culture, ethnicity, religion, and characteristics of the death
9. Avoid analysing or interrupting parents' repeated stories and tears
10. Help to identify and resolve secondary losses, such as the hopes, dreams and expectations the parents had for the deceased child
11. Examine defenses and coping strategies; carefully examine resistance to the grief process
12. Assist in finding sources of continuing support
13. Identify and refer "pathology"
14. Interpret "recovery" for them; correct unrealistic expectations of themselves and of the grief process

Families may value opportunities to meet with the team who have played a role in caring for the baby. Social workers, psychologists and volunteers who have been part of the care giving team can play an important role in caring for the bereaved family.

Organization of perinatal palliative care services

From the above discussion, it is clear that babies with life threatening anomalies and their families have complex care needs. A multi disciplinary team of professionals who are skilled in providing compassionate and competent care will play an important role in improving the care of these infants.

Specialist pediatric palliative care teams must be available for consultation at all centers that provide care for dying children and neonates. These teams may be even available for consultation on the telephone to the primary pediatric teams.

The team members of the pediatric palliative should be- physicians with appropriate training, nurses, rehabilitation therapists, psychologists, child life therapists, chaplains, social workers and play therapists. Access to other professionals like psychiatrists must also be available.

In perinatal care, palliative care teams can be involved in caring for the family from the time of diagnosis of a life threatening anomaly up to the death of the infant and through bereavement. The palliative care team should be involved along with the obstetrician, neonatologist and of the members of the primary care team.

To summarize the above discussion, the palliative care working in the perinatal setting can play an important role in the following areas-

1. Communication, breaking bad news and providing information
2. Providing emotional support to the family
3. Comfort care for the infant
4. Caring for siblings
5. Bereavement support
6. Educating other professionals in palliative care
7. Advocacy for palliative care provision

The ACT organization, UK¹⁷ provides a framework for organizing perinatal palliative care services in an organization. A brief outline of such a service based on the ACT recommendations is given below.

Standard 1

- **Breaking bad news**

Issues- Family-Information – choices, training in care of baby, transport, siblings, contact details for professionals, post-natal care of mother, bereavement support, emotional support, religious and spiritual support

Baby-Symptom control, emergency care plan, baby care needs, registration of birth

- **Planning to go home**

Family-All of the above and plan for emergency situations

Baby- Symptom control, equipment, follow up

Standard 2

- **Ongoing care for the baby and the family**

All the above issues must be addressed and an effective response system must be arranged. Liaising with local medical practitioners and pediatricians is often helpful.

End of life care

Family: Information – choices, sibling involvement, contact details for professionals emotional support, spiritual issues, funeral planning, grandparents, post-natal care of mother, memory box

Baby: Symptom control, cultural/religious issues, allowing a natural death care plan

Bereavement

Family- Support for as long as is necessary

Baby-Funeral, burial/cremation, ceremony/ies following the death of baby, registration of death

Conclusion

Caring for neonates with congenital anomalies is a challenging task. The palliative care approach is likely to benefit the quality of care received by the babies and the families.

Pediatric palliative care is in its infancy in India. Professionals caring for women with fetuses with congenital malformations can liaise with pediatric palliative care specialists in order to improve the care that their patients receive.

Points to Remember

- *Breaking bad news to parents when the fetus is detected to have an anomaly needs skill and compassion.*
- *Counseling parents in the antenatal period must take the prognosis for survival into account.*
- *Physicians must provide adequate information and support to parents so as to help them make appropriate decisions regarding termination of pregnancy.*
- *Parents must receive adequate information about the prognosis of the resuscitation efforts and must receive appropriate emotional support to make decisions.*
- *All babies who need palliative care must be monitored for pain*
- *Social workers, psychologists and volunteers who have been part of the care giving team can play an important role in caring for the bereaved family.*

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CLIPPINGS

Antibiotics for middle-ear infection (acute otitis media) in children

Acute otitis media (AOM) is one of the most common diseases in early infancy and childhood. Antibiotic use for AOM varies from 56% in the Netherlands to 95% in the USA, Canada and Australia. The objective was to assess the effects of antibiotics for children with AOM. Antibiotic treatment led to a statistically significant reduction of children with AOM experiencing pain at two to seven days compared with placebo but since most children (82%) settle spontaneously, about 20 children must be treated to prevent one suffering from ear pain at two to seven days. Additionally, antibiotic treatment led to a statistically significant reduction of tympanic membrane perforations (NNTB 33) and contra lateral AOM episodes (NNTB 11). These benefits must be weighed against the possible harms: for every 14 children treated with antibiotics, one child experienced an adverse event (such as vomiting, diarrhea or rash) that would not have occurred if antibiotics had been withheld. Antibiotics appear to be most useful in children under two years of age with bilateral AOM, or with both AOM and otorrhea. For most other children with mild disease, an expectant observational approach seems justified. We have no trials in populations with higher risks of complications.

Venekamp RP, Sanders S, Glasziou PP, Del Mar CB, Rovers MM. Antibiotics for acute otitis media in children. *Cochrane Database of Systematic Reviews* 2013, Issue 1. Art. No.: CD000219. DOI: 10.1002/14651858.CD000219.pub3. Published Online: January 31, 2013.

NEWS AND NOTES

XI National Conference of Pediatric Rheumatology Society, Gujarat.

Dates: 24th & 25th August, 2013

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SUPPORTIVE AND PALLIATIVE CARE

COMMON PROBLEMS AND THEIR MANAGEMENT IN PEDIATRIC PALLIATIVE CARE

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Abstract: *Today with growing burden of disease and children with terminal illness, there is a pressing need for pediatric palliative care. This is an approach aimed at providing holistic care to children with terminal illnesses and their family. Pediatric palliative care is unique in its approach due to dissimilarity in the presentation of symptoms between children and adults. This review article outlines various salient features of pediatric palliative care which may aid the professionals caring for children with life limiting illnesses.*

Keywords: *Pediatric palliative care, Symptoms.*

Today, due to tremendous advancements in medical science, children with life limiting illness have improved survival. However, approximately 25% of these children would die of their disease after many years. The provision of high-quality, appropriate and effective palliative care for children is a global concern. The spectrum of life threatening illness in children is different from that in adults. Children with advanced cancer present with varied symptoms who are often inadequately treated. This could be because of multiple reasons and few among them being-lack of self-reporting by children, non-availability of standard assessment tools, reluctance from the side of the physicians to actively seek symptoms. There is dearth of literature regarding symptoms of children dying of cancer. The more

common symptoms experienced include, lack of energy, pain, fatigue, nausea, vomiting, dyspnea, anorexia and psychological symptoms.¹

Most children who die are cared for by general pediatricians or by pediatric specialists in a child's particular disease. Since training in the care of dying children is still not commonly available, this chapter hopes to help those who encounter this situation infrequently. This chapter will consider common symptoms other than pain, which are associated with a variety of life limiting illnesses.

When dealing with the physical symptoms of the child with terminal illness, it may be important to remember that the goal is to maximize comfort and quality of life, while weighing the possible side effects and benefits of a given treatment. The initial step is to take a careful history and make a thorough assessment. In contrast to pain, formal assessment tools for other symptoms in children are not available. Thus it is the child's own description of his / her symptom, which depends on child's age and understanding and the parent's observations and opinion. It is very essential at this stage to consider the quantum of distress the symptom causes to the child and how much does this interfere with child's and family's daily living. If the cause can be identified it could be well treated, although often at this stage it will not be reversible. Thus management at this stage should include drug treatment, nursing care and psychological support both for the child and the family.

Oral problems

Oral health is an important contributor to the health and well-being of any patient. Terminally ill children with debility and lowered immunity often develop oral problems. Dry mouth, dry lips, oral mucositis and oral candidiasis are the commonest problems in palliative care setting (Table.I). Contributing factors include general debility, reduced immunity, poor oral intake, chemotherapy and radiation. Preventive measures include:

1. Brushing of teeth at least twice a day, remove dentures when not in use, soak dentures in mild antiseptic overnight and clean tongue: gentle brushing, irrigate mouth with warm saline.

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Table I. Management of common oral problems

Syndrome	Management
Xerostomia ²	<p>Salivary gland stimulant: Sugarless gums and candies Anticholinergics: Pilocarpine</p> <p>Salivary substitutes: Topical products containing olive oil, betaine and xylitol</p> <p>Newer products with enzyme systems such as lactoperoxidase, lysozyme and glucose oxidase (e.g. Biotène® Oralbalance dry mouth gel) offer potential antimicrobial and moisturizing benefits.</p>
Oral mucositis ³	<p>Treat infection</p> <p>Pain: Local anesthetics such as lidocaine, diphenhydramine, benzocaine lozenges</p> <p>Liquid oral or parenteral opioids may be required for adequate pain management</p>
Oral candidiasis ⁴	<p>Topical</p> <p>Nystatin suspension (“swish and swallow”)</p> <p>Clotrimazole troches are a reasonable first line therapy for patients in the palliative care.</p> <p>Fluconazole or newer generation antifungals are recommended (if above fails).</p> <p>Systemic: Fluconazole/Itraconazole/Amphotericin</p>

- Rinse mouth twice daily using antiseptic or antimicrobial mouthwashes (alcohol-free). Mouth washes such as 0.12% chlorhexidine gluconate, povidone-iodine, lemon and glycerine or even effervescent solution- soda water twice daily may be effective in preventing dental caries and oral infections.
- Aphthous ulcer: use tetracycline suspension mouth wash or apply Orabase gel
- Malignant ulcers: Antibiotics to cover anaerobic organisms. A course of metronidazole can be useful for malodorous fungating and ulcerating lesions.

Nausea and vomiting

Nausea and vomiting are relatively common unpleasant symptoms in children receiving palliative care. Causes of nausea and vomiting may include raised intracranial tension, metabolic disturbances, side effects of drugs, radiation and external pressure on the bowel. The control and neurophysiology of nausea and vomiting is complex and can be induced by a variety of stimuli. Fig.1 depicts the neurophysiology of nausea and vomiting.

A thorough knowledge of the etiology and pathophysiology of the condition is crucial. The optimal management of nausea and vomiting is based on ongoing assessment and historical documentation including biochemical analysis. The choice of anti- emetic depends

on the underlying cause and the receptor being stimulated for instance, haloperidol is the antiemetic of choice for Morphine induced nausea and vomiting (Table II). In situations where the cause is not immediately established, universal antiemetic may be used, which include metoclopramide and domperidone in standard doses.⁵ Although the role of ondansetron in palliative care, even in adults, has not been defined, studies suggest that it can be very helpful when vomiting is severe and resistant to other antiemetics.⁶ Table II summarizes the anti-emetics of choice in specific conditions.⁷

Constipation

Terminally ill children have many problems which can contribute to constipation. These include poor fluid and dietary intake, lack of mobility, weakness and wasting and the use of drugs like opioids, phenothiazines and anticholinergics. Fortunately in children, this is often balanced due to their younger and fitter bowels. If parents are aware of the possibility of constipation developing and keep note of bowel action, then simple measures and mild laxatives can be administered and severe problems can be avoided.

Pathophysiology of constipation: Constipation is frequently multifactorial in origin and result from systemic or neurologic disorders as well as from medication. Causes are classified into:^{7,8}

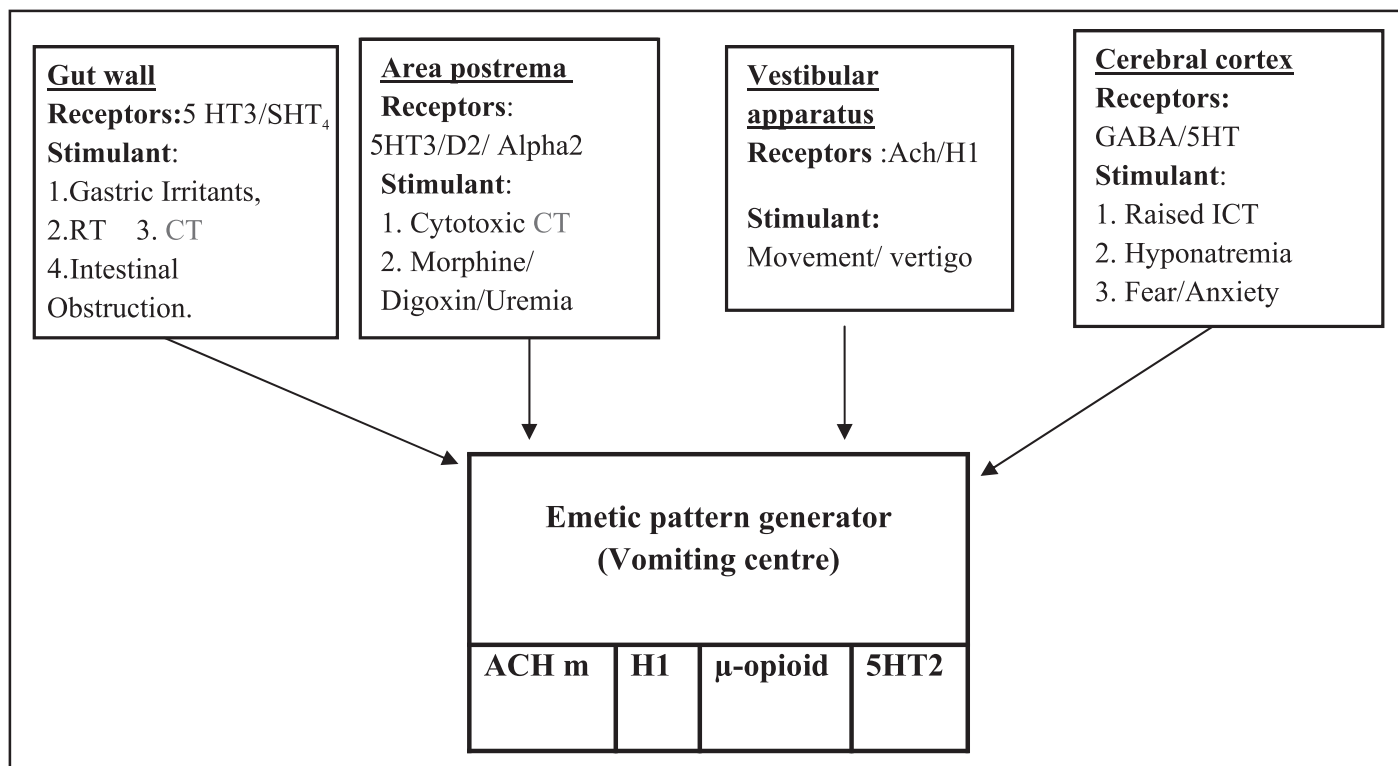


Fig.1. Neurophysiology of nausea and vomiting

1. **Normal-transit constipation:** The stool traverses at a normal rate and frequency is normal, but the patient perceives difficulty in evacuation. This may be due to abdominal bloating, psychological distress, increased rectal compliance and decreased rectal sensation.
2. **Slow-transit constipation:** Infrequent urge to defecate, bloating, abdominal pain or discomfort are often symptoms associated with this condition. Patients with minimal disorder respond well to high fiber and laxatives. In non responsive patients, there is often delayed emptying of the proximal colon and relatively few high amplitude peristaltic contractions after meal.
3. **Defecation disorder:** This is due to dysfunction of the pelvic floor or anal sphincter. Failure of the rectum to empty effectively may be due to an inability to coordinate the abdominal, rectoanal, pelvic floor muscles during defecation and can be functional or organic. Functional defecatory dysfunction can be avoided by prolonged avoidance of pain for example anal fissures.

Causes of constipation: 1. **Primary:** Reduced fiber, fluid, mobility and privacy for evacuation 2. **Secondary:** Structural causes such as tumor, neural infiltration of tumor, electrolytes (hypercalcemia for certain types of cancer), endocrine - hypothyroidism and diabetes mellitus, neurological, pain, para-neoplastic syndrome

3. Medications that can cause/exacerbate constipation: Opioids, morphine, codeine (95% of patients taking opioids), anticholinergics: tricyclic antidepressants, antispasmodics, sympathomimetics: ephedrine, terbutaline, others like lithium, verapamil, bismuth, iron, aluminum (antacids) calcium salts - calcium carbonate, barium sulfate, diuretics, ferrous sulphate, antacids, 5HT₃ antagonist, vinca alkaloids, NSAID like ibuprofen. Medical management of constipation is summarised in Table.III.

Diarrhea

This is a distressing symptom to families and cause considerable practical problems. Pharmacological approaches to relieve the symptoms need to be given alongside investigations to identify the cause.

Causes: Multiple factors can cause diarrhea: villous atrophy due to immune mediated HIV enteropathy, necrotizing enterocolitis resulting from chronic intestinal infection, overuse of laxatives, severe (often a neglected) constipation and fecal impaction can also cause diarrhea as backed- up, liquefied stool may be all that the patient can pass ('overflow diarrhea'), partial intestinal obstruction, chemotherapy and radiation enteritis.

Management: Pharmacological approaches to relieve the symptom need to be given alongside investigations to identify the cause. Oral loperamide is often effective in treating the cause, if not then, oral morphine may be added.⁹

Table II Reversible causes of vomiting and their management

Cause of vomiting	Treatment of underlying cause	Anti-emetics of choice
Gastric irritation	Identify the underlying etiology and modify the management	1. Ondansetron+dexamethasone 2. Haloperidol 3. Levomepromazine\$
Vestibular Disturbance		1. Cyclizine*/ Hyoscine patch/ Scopalamine 2. Antihistaminics 3. Cinnarazine
Opioid induced vomiting	Prophylactic metoclopramide/Laxatives	1. Haloperidol 2. Metoclopramide/Ondansetron
Hypercalcemia	Rehydration Bisphosphonates	1. Haloperidol#* 2. Setrons@ 3. Levomepromazine\$
Raised intracranial pressure	Dexamethasone	1. Cyclizine 2. Levomepromazine\$
Systemic infection	Treat infection(Antibiotics, antivirals, antifungals)	1. Haloperidol#* 2. Setrons@ 3. Levomepromazine\$
Uremia	Dopamine/Histamine Antagonist Correct electrolyte Corticosteroids	1. Haloperidol#* 2. Setrons@ 3. Levomepromazine
Anxiety	Explanation and reassurance Anxiolytics Dexamethasone	1. Cyclizine
Partial Bowel obstruction/ Gastric stasis		1. Metoclopramide/Domperidone 2. Haloperidol 3. Dexamethasone/Levomepromazine
Complete Obstruction		1. Cyclizine 2. Levomepromazine 3. Octreotide

1.First anti-emetic of choice, 2. Second Antiemetic of choice, 3.Third antiemetic of choice*Drug of Choice, CTZ- chemoreceptor trigger zone, #-D2 blocker, @setrons- ondansetron, granisetron, palonosetron, \$-5HT-2 blocker, VC-vomiting centers

If diarrhea is severe, morphine can be given subcutaneously. Octreotide, a somatostatin analogue, has been successfully used in adults, may also have a role in refractory cases in

children. Consideration of practical issues like skin hygiene and sanitation of the home environment are also equally essential in care of children.

Table.III. Pharmacological treatment of constipation⁷

Class of drug	Drugs
Osmotic laxative- Produces osmotic effect in colon, that results in distention and promotes peristalsis	Lactulose, sorbitol, magnesium hydroxide, sodium phosphate(>5 years, C/I in renal failure)
Colonic Stimulants- Promote peristalsis	Bisacodyl, senna, castor oil
Lubricant- Soften stools and decrease water absorption from GI	Mineral Oil(Avoid by mouth as it can cause aspiration pneumonia and depletion of A,D,E,K vitamins.)
Bulk forming agents- Absorb water in intestine to form viscous liquid that promote peristalsis and reduces transit time	Psyllium (mix with water to prevent choking, low fluid intake may cause impaction) Methylcellulose (low fluid intake can cause impaction)
Emollient stool softener- Helps keep stools soft for easy natural passage	Sodium docusate(<3 years) Calcium docusate(3-6years)

Anorexia

Anorexia and weakness are two of the commonest symptoms described by patients with advanced cancer and AIDS. They often pose a difficult problem for patients and care takers. Failure to eat is often perceived as a sign of giving up by the patient while caretakers constantly feel that if the patient eats he will get better.

Mechanism: Anorexia is a multifactorial process and the exact cause and mechanism remains unknown. It is speculated that pro-inflammatory cytokine - IL-8, TNF play a role.¹⁰

Causes: Multiple factors cause anorexia like oral mucositis, xerostomia, oral candidiasis, chemotherapy, radiation, depression and infection.

Management: Identify the appropriate cause and correct it. Depression is often overlooked, which needs assessment and appropriate referral to specialist.⁹ Assess nutritional status and refer for interventional management like percutaneous endoscopic gastrostomy tubes and nasogastric feeds. However parenteral or forced and expensive enteral nutrition therapy should be discouraged in cachectic and terminally ill patients.

Drug therapy

Drugs with known benefits

1. High dose progestins
2. Megesterol acetate¹¹

Drugs with doubtful or controversial benefits

1. Corticosteroids¹²
2. Appetite stimulant like cyproheptadine¹³
3. Dronabinol

Dyspnea

Breathlessness is one of the most frightening and distressing symptoms, often accompanied by anxiety in both children and parents. Some children may experience severe panic attack and become convinced that they are about to die. Dyspnea occurs in 40-65% of children with malignant conditions and there is evidence that control of dyspnea may be less effective than that of pain in palliative care.¹⁴

Though the precise cause is not known. Some of the causes can be classified into three categories:¹⁵

1. An increase in the respiratory efforts to overcome a certain load: obstructive or restrictive lung disease, pleural effusion.
2. An increase in the proportion of respiratory muscles required to maintain a normal workload,
3. Increase in the ventilatory requirements like hypoxemia, hypercapnia, metabolic acidosis, anemia.

Management : The first principle in the management; include identifying the putative factor and treating it.

This may not be possible or appropriate in the palliative care setting and needs to be considered on an individual basis.

Oxygen may be helpful if there is hypoxia and cyanosis or sudden episode of hyperventilation.

Non pharmacological Support

Reposition (upright) the patient to relieve distress, avoid compression of abdomen or chest when positioning.

Offer psychosocial support and/or counseling and breathing techniques.

Pharmacological support: Opioids: In one of the randomized trials, opioid (morphine) was administered at one quarter the regular 4 hour dose was found to sufficiently reduce respiratory distress and tachypnoea.¹⁶ Nebulised morphine is effective in some patients with fewer systemic side effects. Caution is needed in its use due to limited experience and potential for causing bronchospasm. **Corticosteroids** like dexamethasone are particularly indicated in the presence of bronchial obstruction, SVC or lymphangitic carcinomatosis. **Benzodiazepines** are used in combination with opioids due to its anxiolytic and sedative effects. They also reduce seizures and relax muscles. **Nebulisation** with bronchodilators with steroids with or without salbutamol / ipratropium bromide.¹⁵

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NEWS AND NOTES

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SUPPORTIVE AND PALLIATIVE CARE

PAIN MANAGEMENT IN PALLIATIVE CARE

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Abstract: *Palliative care for children is the active total care of the child's body, mind and spirit and also involves giving support to the family. It begins when illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease. Health providers must evaluate and alleviate a child's physical, psychological and social distress. This article narrates an overview of paediatric pain in brief.*

Keywords: *Palliative care, Pediatric pain, Pain assessment, Children.*

The emotional and mental trauma that parents undergo after diagnosis of a malignancy in their child is enormous. In addition, if the child is in pain the emotional burden increases, making the parents feel more helpless. Most parents of children with cancer have dual primary goals: a primary cancer-directed goal of cure and a primary comfort-related goal for lessening suffering.¹ Palliative care in pediatric setting should begin right from the diagnosis of the illness and continue through the course of the disease.

Appropriate supportive measures to address the other physical and psychosocial problems that the child and family are facing should also be instituted. WHO definition of palliative care appropriate for children and their families is: "Palliative care for children is the active total care of the child's body, mind and spirit, and also involves giving support to the family. It begins when illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease. Health providers must evaluate and alleviate a child's physical, psychological, and social distress."²

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An American Academy of Pediatrics (AAP) policy statement recommends that "all general and subspecialty pediatricians, family physicians, pain specialists, and pediatric surgeons need to become familiar and comfortable with the provision of palliative care to children."³

The incidence of cancer is increasing worldwide. Unfortunately in India due to ignorance or lack of access to medical facilities or financial constraints the number of children succumbing to cancers is still high, despite the advances made in treatment of various malignancies worldwide.

So, in Indian scenario, the relief of pain and institution of palliative measures becomes all the more important due to the higher morbidity and mortality associated with paediatric malignancies. Palliative care should be instituted with appropriate drugs and other supportive measures as pain in a child may make the child noncompliant to treatment and the parents to lose their trust in the treating personnel⁴. Inadequate relief of pain has been found to lead onto post traumatic stress disorder, depression and stress even years after treatment.²

Clinicians often have misconceptions regarding the ability of children to perceive pain and to understand the nature and extent of pain completely, though it has been shown that even a 26 week neonate can appreciate and experience pain.⁵ Pain is often undertreated in children due to fear of addiction to opioids and misconceptions regarding opioid usage and ignorance regarding pharmacodynamics which may lead on to inadequate and improper usage. Health professionals dealing with pediatric malignancies often lack knowledge regarding simple cognitive, behavioural and supportive techniques for pain management.

Pathophysiology

Pain can be broadly classified into nociceptive, neuropathic and mixed types. The mechanisms from which pain arises are numerous and a detailed review is beyond the scope of this article.

Major types of pain in children are shown in Table.I.

Table.I.Pain in children

Caused by disease	Caused by anti cancer treatment
Tumour involvement of bone	Post operative pain
Tumour involvement of soft tissue	Radiation induced skin changes
Tumour involvement of viscera	Gastritis from repeated vomiting
Tumour involvement of CNS or PNS	Neuropathy post chemotherapy
	Mucositis
	Venepuncture or injections
	Diagnostic procedures like lumbar puncture, biopsy etc

Pain management

In developed countries cancer pain in children is therapy or diagnostic procedure related whereas in developing countries the pain is mainly disease related.²

The management of pain in children is complex because of the inability to assess the presence of pain (in infants) and children cannot fully express the degree of pain and its nature and site⁴, but once relieved of pain children immediately express it by resuming their routine activities. Children are dependent on their caregivers for recognising their pain and instituting treatment.

According to WHO guidelines pain evaluation can be done by **ABCDE** steps:

Assess: Always assess children for pain though they may not be able to express it verbally.

Body: Physical examination of the child to find out area of pain is essential. The area of pain is usually indicated by grimacing, crying, contracture, rigidity, etc. The relationship of disease and area of pain should also be established.

Context: Consider the impact of the family, environment, health care, etc on child’s pain and its management

Document: During each visit of the child, document the pain score using pain assessment scores. Use age appropriate scores for pain score documentation

Evaluate: Regularly evaluate the effectiveness of therapy and modify accordingly.

Another comprehensive method of pain assessment is **QUEST**⁶

Question the child.

Use pain rating scales.

Evaluate behaviour and physiologic changes.

Secure parents’ involvement.

Take cause of pain into account.

Take action and evaluate results.

Pain assessment scores

A variety of scores have been used by clinicians for the evaluation of pain and denoting the degree of severity of pain (Fig.1 & Fig.2).

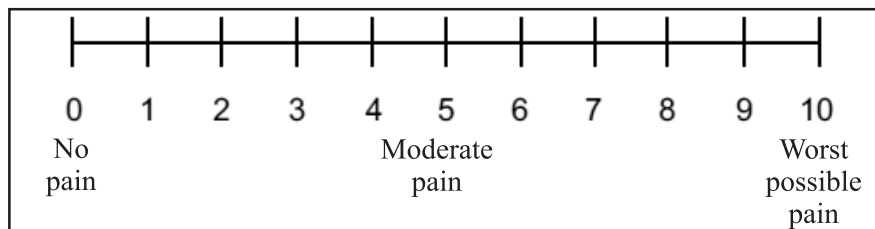


Fig.1. 0-10 Numeric Pain Intensity Scale⁷

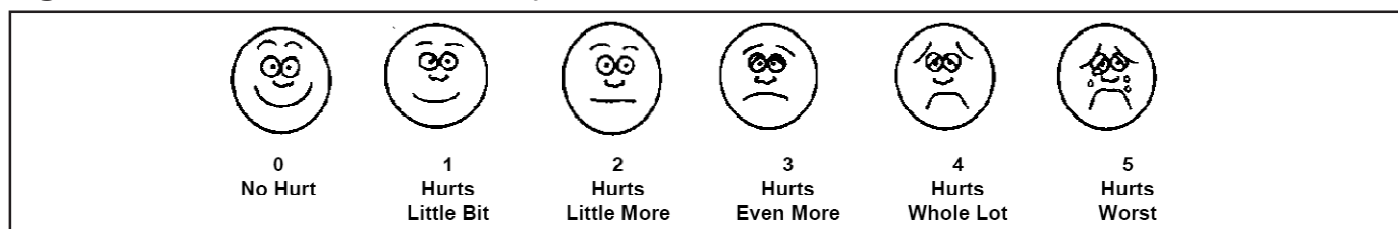


Fig.2.Wong-Baker FACES Pain Rating Scale

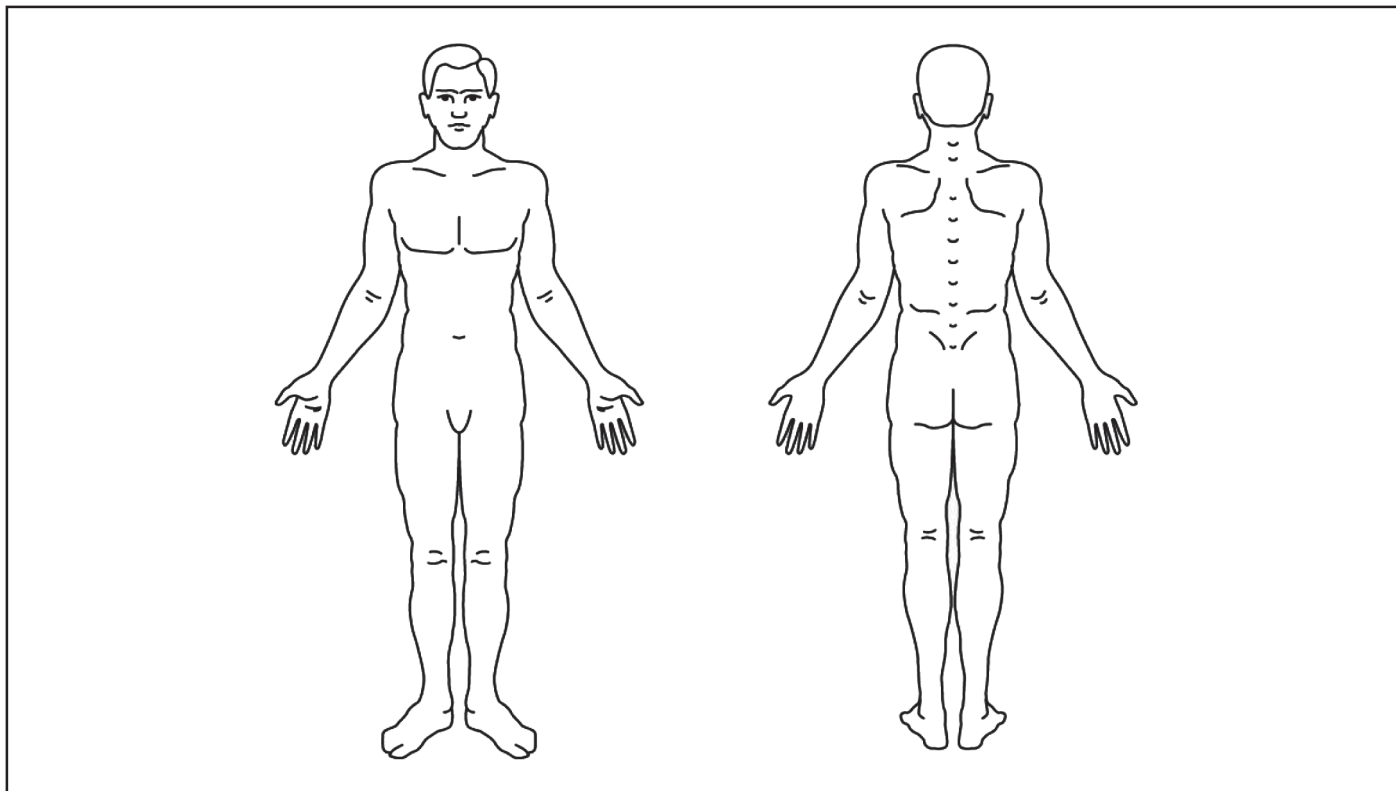


Fig.3. Pain localisation

Picture of a human body can also be shown so as to make the child identify the area of pain (Fig.3)

Drug therapy

Ideally WHO principles should be used in managing pain in children

By the ladder: Start from milder analgesics and proceed to stronger opioids; to start with paracetamol then add codeine or morphine as needed (Fig.4). Move up the ladder, if adequate pain relief is not obtained adjuvants should be added as needed.^{8,9}

By the clock: Medications should be given regularly even if pain is not present and not wait for onset of pain to give medications. By prn technique of drug administration the child might be in pain and tends to fear pain, moreover higher dose of drugs are needed to control pain once it arises. Extra doses of medications can be used to control break through episodes of pain.

By oral route: Prefer the least invasive route for giving medications, preferably orally as tablets or elixirs.

By the child: Patient comfort and preferences should be kept in mind while starting and adjusting analgesics. Any side effects should be promptly addressed.

Drugs for pain relief

NSAIDs and paracetamol

Non-steroidal anti-inflammatory drugs (NSAIDs) have

analgesic, anti-pyretic and except for paracetamol-anti-inflammatory activity. NSAIDs act peripherally to provide their analgesic effect by interfering with the synthesis of prostaglandin, through the inhibition of two isoenzymes of cyclooxygenase (COX); COX-1 and COX-2. The COX-1 isoform is expressed primarily in kidney, gastrointestinal tract and on platelets. The COX-2 isoform is found in low levels in tissues, but is induced during inflammation. Data for safety regarding use of COX-2 inhibitors in children is inadequate though celecoxib has been used in treatment of Ewings sarcoma due to its antiangiogenic properties.^{10,11}

The side effects of non selective NSAIDs include decreased platelet aggregation, gastric irritation, and potential for renal toxicity with long term use. Children with low platelet counts or who are neutropenic should be monitored carefully when taking non selective NSAIDs for pain relief. Hepatic toxicity can occur with high doses of paracetamol (i.e., >75 mg/kg/day or 4g/day) though otherwise well tolerated. If pain increases or persists beyond the maximum dosage of a NSAID, switching to a opioid would be the next step in managing a child's pain. NSAIDs are limited to a ceiling dose where increasing the dose only produces toxicity and no pain relief (Table II).

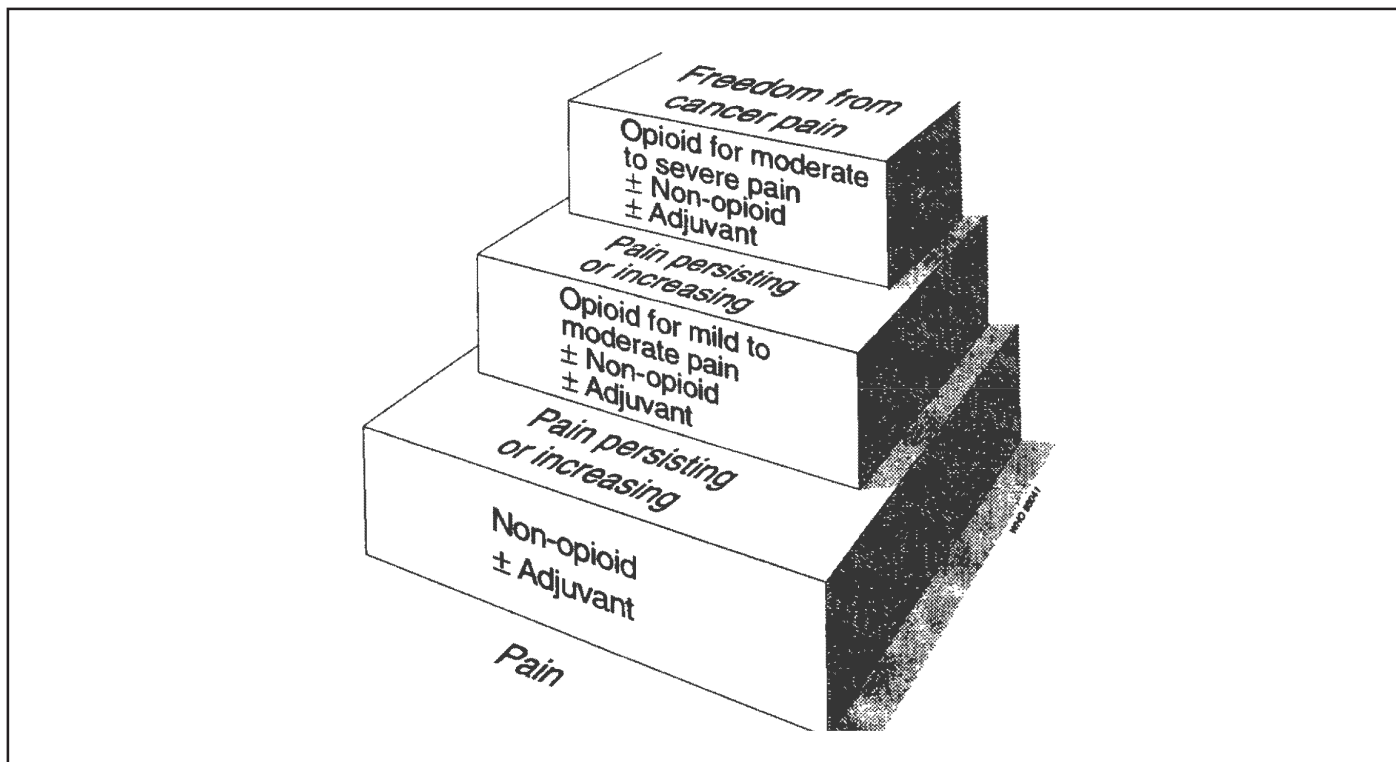


Fig.4. Approach to drug therapy for pain

Table.II. Drugs for pain therapy and their dosage

Drug Type	Typical Starting Dose
Paracetamol	10 to 15 mg/kg/dose q 4 hr PO to a max of 650 mg/dose
Ibuprofen	10 mg/kg/dose to a max single dose of 800 mg q 6 to 8 hr PO
Diclofenac sodium	1 to 1.5 mg/kg/dose to a max single dose of 75 mg q 8 to 12hr PO
Naproxen	5 to 7.5 mg/kg/dose to a max of 500 mg/dose q 12 hr PO

Opioids

Opioids bind with receptors (Mu, Kappa, Delta) in the CNS and peripheral tissues to provide analgesic effects

Mu-agonist drugs are the most commonly used class of opioids and include drugs such as morphine, fentanyl, and codeine. No ceiling dose has been observed for their analgesia with dose limitation being side effects.¹²

Depending on the opioid, they can be given orally, rectally, as subcutaneous or intravenous infusions, intramuscularly, transe dermally, and directly into the CNS via epidural/caudal/intrathecal injection.

The most common side effects of opioids analgesia in children are constipation, sedation, pruritis, and nausea/vomiting. Respiratory depression, although the most

frequently cited concern of health care providers, is a relatively rare occurrence.

Codeine: Codeine is commonly used for pain relief usually in combination with non opioids, but switch over to stronger opioids if inadequate relief is obtained. In children less than 6 months half to one third the recommended dose should be started before titrating upward. If inadequate relief, switch over to stronger opioids. Parenteral administration is not recommended.

Morphine: Morphine is one of the commonest and cheapest form to relieve cancer pain. But sadly it is still not available in many parts of the world. Morphine is available as immediate release and sustained release form apart from injectable form. Morphine syrup is still not available in India, though indigenous preparations are available at a few centres.

Table.III.Dosage guidelines for opioids^{2,4}

Drug	Oral starting doses	Dosage forms	Starting DosesIV	IV toPO
Codeine	0.5 to 1 mg/kg q 4- 6 hr; max: 60 mg/dose	Tablet, as sulfate: 30 mg Liquid: 3 mg/mL	N/A	N/A
Morphine	0.3 to 0.6 mg/kg/dose every 8 -12 hr for sustained release 0.2 to 0.5 mg/kg/dose q 4to 6 hr prn for solution of instant release tablets	Injection: 2 mg/mL, 5 mg/mL, 10 mg/mL, 15 mg/mL Injection, preservative free: 1 mg/mL Solution: 2 mg/mL Tablet: 15 mg (instant release) Tablet, controlled release: 15 mg, 30 mg, 60 mg, 100 mg, 200 mg	0.1 mg/kg/ dose 0.1 to 0.2mg/kg/dose q 2 to 4 hr; max: 15mg/dose	10 mg IV to 30 mg PO
Hydromorphone	0.03 to 0.08 mg/kg/dose PO q 4 to 6 hr; max: 5 mg/dose	Injection: 1, 2, 3 and 4 mg/mL Tablet: 2 mg, 4 mg Syrup: Hydromorphone 1mg and guaphenesin 100 mg/5 mL Suppository: 3 mg	15 mcg/kg IV q 4 to 6 hr; max: 2mg/dose	1.5 mgIV = 7.5 mgPO
Methadone	0.1 to 0.2 mg/kg q 4 to 12hr; max: 10 mg/ dose	Tablet: 5 mg, 10 mg Solution: 1 mg/mL Concentrate: 10 mg/ml Injection: 10 mg/mL	0.1mg/kg IV q 4- 12 hr; max: 10mg	10 mgIV = 20 mgPO
Fentanyl	Lozenge: < 15 kg: contraindicated; > 2 years (15 kg to 40 kg):5 to 15 mcg/kg; max:dose 400 mcg > 40kg 5 mcg/kg; max:dose of 400 mcg	Lozenge: 100 mcg, 200 mcg, 300 mcg, 400 mcg Patch: 25 mcg/hr, 50 mcg/hr, 75mcg/hr, 100mcg/hr Injection: 50 mcg/ml	1 to 2mcg/kg/dose; max: 50 mcg/dose Continuous IV infusion: 1mcg/kg/hr	NA

Morphine should be started in children after giving a be given at one third to one half the recommended dosage in infants less than six months old.

Morphine doses need to be titrated to achieve adequate pain control and pain relief should be balanced with side effects. Initial dosage titration is preferred with immediate release preparations and once dosage stability is achieved patient can be switched over to sustained release preparation as less frequent dosing schedule is required. (Table.III) Crushing of sustained release preparations should be discouraged as the drug loses its potency.

Dosage modification should be done if co- existing renal or hepatic failure exists or if patient is debilitated. Oral to parenteral ratio is 3:1. Parents should be educated regarding the side effects of drug prior to starting morphine.

Hydromorphone: Six times more potent orally and eight times more potent parenterally than morphine.

Though not available in India, it is an alternative if side effects of morphine are unbearable. Hydromorphone is similar to morphine in its pharmacokinetics, efficacy and toxicity.

Methadone: Synthetic long acting derivative of morphine which is recommended if side effects develop to the above two drugs. Due to its long duration the child should be observed for a few days for signs of drug toxicity. Oral to parenteral ratio is 2:1 and dose reduction should be considered if child is debilitated with coexisting renal or liver failure.

Fentanyl: Short acting opioid used for acute pain relief, mainly procedure related. Fentanyl is administered as either

Table.IV. Side effects of opioids and their treatment

Side effects	Treatment (Drugs)	Non pharmacologic techniques
Constipation	Senna and docusate sodium: Tablet: 2 to 6 years= Start: ½ tablet once a day; max: 1 tablet twice a day 6 to 12 years =Start 1 tablet once a day; max:2 tablets twice a day > 12 years= Start 2 tablets once a day; max: 4tablets twice a day Liquid:1 month to 1 year: 1.25 to 5 mL q hs 1 to 5 years = 2.5 to 5 mL q hs 5 to 15 years = 5 to 10 mL q hs > 15 years = 10 to 25 mL q hs Bisacodyl: PO or PR 3 to 12 years 5 mg/dose/day > 12 years 10 to 15 mg/dose/day Lactulose: 7.5 mL/day after breakfast	Increase water intake Prune juice, bran cereal, vegetables
Vomiting	Promethazine: 0.5 mg/kg q 4 to 6 hr; max: 25mg/dose Ondansetron: 0.1 to 0.15 mg/kg IV or PO q 4 hr;max: 8 mg/dose Granisetron: 10 to 40 mcg/kg q 2 to 4 hr; max: 1 mg/dose Droperidol: 0.05 to 0.06 mg/kg IV q 6 hr	Imagery, relaxation, Deep, slowbreathing
Pruritis	Diphenhydramine: 1 mg/kg IV / PO q 4 to 6 hr prn; max: 25 mg/dose Hydroxyzine: 0.6 mg/kg/dose PO q 6 hr; max: 50 mg/dose Naloxone: 0.5 mcg/kg/hr continuous infusion (diluted in a solution of 0.1 mg of naloxone per 10 ml of saline) Butorphanol: 0.3 to 0.5 mg/kg IV (use cautiously in opioid tolerant children, may cause withdrawal symptoms); max: 2 mg/dose because mixed agonist/antagonist	Oatmeal baths, good hygiene. Exclude other causes of itching Change opioids
Respiratory depression: Mild—moderate	Hold dose of opioid Reduce subsequent doses by 25- 50 %	Arouse gently, give O ₂ , deep breath
Respiratory depression: Severe	Naloxone: During disease pain management: 0.5 mcg/kg in 2 minute increments until breathing improves Reduce opioid dose if possible Consider opioid switch During Sedation for Procedures: Naloxone 5 to 10 mcg/kg until breathing improves Reduce opioid dose Switch opioids	Oxygen supplementation, bag and mask ventilation if indicated
Dysphoria/ confusion/ hallucinations	Evaluate medications, eliminate adjuvant medications with CNS effects as symptoms allow. Consider opioids switch if possible Haloperidol : 0.05 to 0.15 mg/kg/daydivided in 2 to 3 doses; max: 2 to 4 mg/day	Rule out other physiologic causes

bolus or as infusion as an intravenous preparation. Fentanyl is also available as patches, lozenges as well as transmucosal forms as lollipops. The more rapidly acting congeners of fentanyl, sufentanyl, remifentanyl, alfentanyl, etc are also available⁴. Transdermal fentanyl is also available which can be used in patients who cannot tolerate morphine due to side effects. Transdermal fentanyl should always be considered after analgesic titration has been done with oral or intravenous opioids, as its onset of action is

12- 16 hrs and elimination is 21 hrs. Fentanyl patches can be used in children older than 12 years weighing > 50 kg. Patches are preferably avoided in situations requiring acute pain relief. Limiting factor for patches are high costs.

Opioid side effects

Side effects should always be anticipated on initiating opioids and parents should be educated so that they promptly report if any occurs(Table IV).^{2,4}

Table.V.Adjuvants in pain therapy

Category/Drug	Dosage	Indication	Comments
Anti depressants			
Amitriptyline	0.2 to 0.5mg/kg PO HS. Titrate upward by 0.25mg/kg every 5 to 7days as needed Available in 10 mg and 25 mg tablets Usual starting dose is 10 to 25 mg HS	Continuous neuropathic pain with burning, aching, dyesthesia with insomnia	Provides analgesia by blocking reuptake of serotonin and norepinephrine possibly slowing transmission of pain signals. Helps in pain related to insomnia and depression (use nortriptyline if patient is over-sedated) Analgesic effects seen earlier than antidepressant effects Side effects include dry mouth,constipation, urinary retention
Nortriptyline	0.2 to 1.0 mg/kg PO a.m. or bid. Titrate up by 0.5 mg q 5 to 7 days Max: 25 mg/dose	Neuropathic pain as above without insomnia	
Anti convulsants			
Gabapentin	5 mg/kg PO at bedtime Increase to bid on day 2, tid on day 3 Max: 300 mg/day	Neuropathic pain	Mechanism of action unknown. Side effects include sedation, ataxia, nystagmus, dizziness
Carbamazepine	2 mg/ kg BD increased to 10- 20 mg/ kg in 2- 3 divided doses	Neuropathic pain, Peripheral neuropathies Phantom limb pain	Analgesic effect as amitriptyline. Monitor blood levels for toxicity. Side effects include pancytopenia, ataxia, and GI irritation
Corticosteroids			
Dexamethasone	Cerebral edema : 1 to 2 mg/kg load then 1 to 1.5 mg/kg/day divided every 6 hr. Anti-inflammatory: 0.08 to 0.3 mg/kg/day divided 6 to 12 hrly	Pain from increased intracranial pressure Bony metastasis. Spinal cord /nerve compression	Side effects include edema, gastrointestinal irritation, increased weight, acne. Use gastro protecting drugs such as H2 blockers (ranitidine) or proton pump inhibitors such as omeprazole for long-term administration of steroids or NSAIDs in end-stage cancer with bony pain

Adjuvants

Adjuvants can be used at any point of the WHO ladder for better pain relief (Table.V)⁴

Tapering opioids

When a patient has adequate pain relief then consider reducing opioids. If patient has received the opioid for more than 3 weeks, taper the medication rather than abruptly stopping. The opioid should be decreased by 20% every other day. If the patient has signs and symptoms of

withdrawal (flu-like symptoms, abdominal cramping, diarrhea), the dose may need to be decreased in smaller amounts over a longer period of time.^{2,4}

Procedure related pain

Adequate analgesia in the form of immediate onset, short acting analgesics should be given as paediatric cancer patients perceive procedures as extremely painful.^{13,14} Procedure should be explained to parents and patient and skilled personnel should perform the procedure in a pleasant environment.¹⁵

Non pharmacological therapy

These have to be implemented along with drug treatments for optimal management of pain. They are only intended to supplement but not replace drug therapies. These therapies require the participation of the caregiver along with the patient.

Supportive therapy: Promotes psychosocial support of the patient. Family participation is essential for this mode of therapy.

Cognitive therapy: Influences the thoughts and images of the child. Parents are the ideal providers of this therapy as they know the preferences of the child. This therapy aims to distract the child’s mind from the pain.

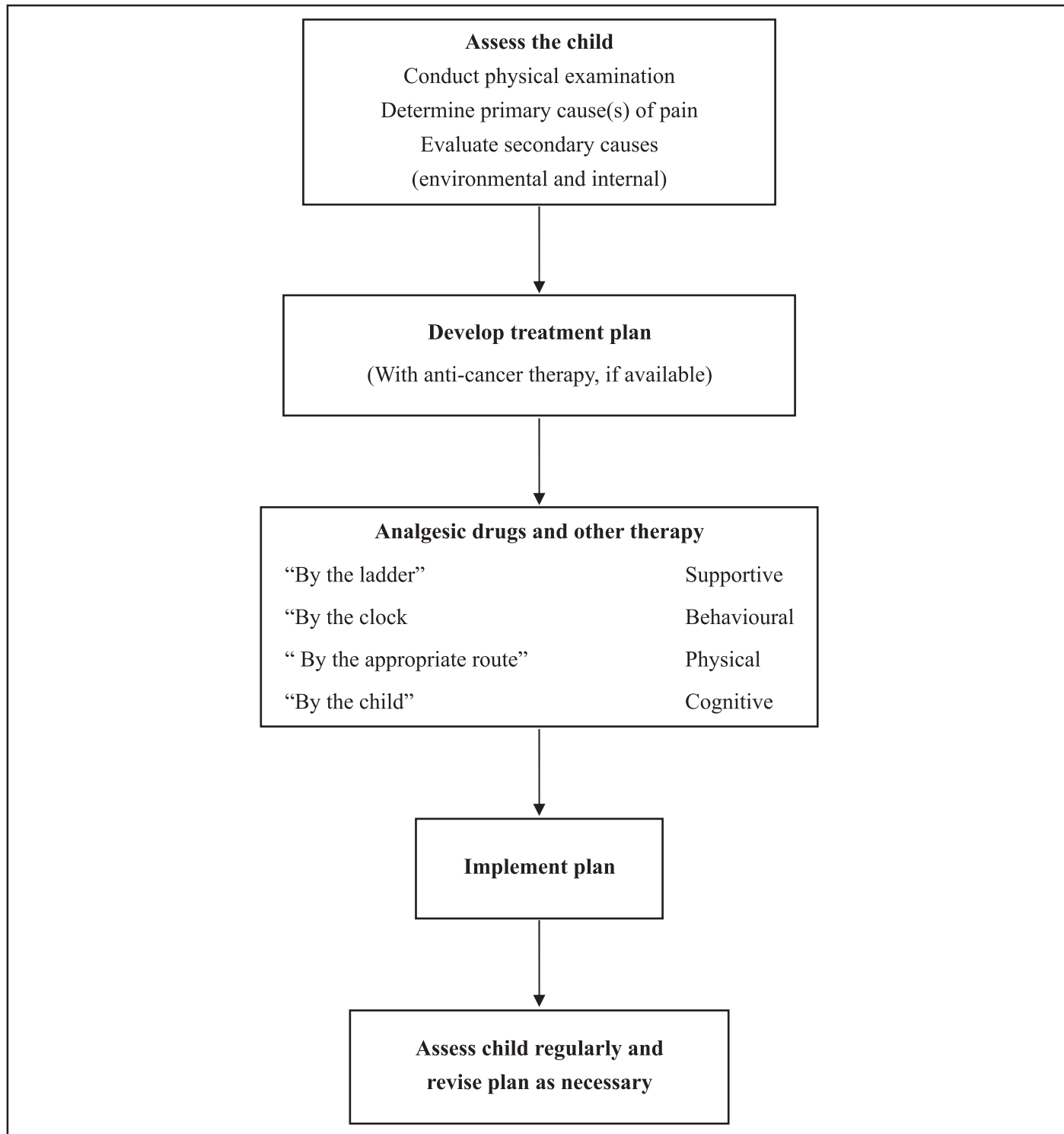


Fig. 5. Approach to a child with pain, as a part of palliative care

Imagery: Imagining a pleasant experience helps divert the child's mind from a past unpleasant experience. Colours, sounds, smell, stories, etc can be used to remember past pleasant incidents.

Behavioural methods-Deep breathing: Simple and effective technique for relaxing the body. Younger children can perform this by blowing bubbles or party blowers and older children can do deep breathing by counting numbers.

Progressive relaxation: Sequential tensing of muscles while lying down combined with deep breathing helps relax the body and helps in reducing anticipatory anxiety and nausea and vomiting. This method is useful in adolescents.

Physical methods-Touch: Physical and psychologically appropriate touch constitutes an important form of therapy, especially in toddlers. Hugging, rocking, swaddling, stroking, caressing and massaging are comforting to the child. Hot packs can be applied for the relief of muscle spasm likewise icepacks can be used to reduce post procedure pain.

Transcutaneous electrical nerve stimulation (TENS): Electrodes placed on the skin help in delivering cutaneous stimulation to large diameter fibres reducing pain transmission at the spinal level. This device can be operated at home after simple instructions.

The entire approach to a child with pain is shown in Fig.5 in summary.²

Points to Remember

- *Palliative care in pediatric setting should begin right from the diagnosis of the illness and continue through the course of the disease.*
- *Palliative care should be instituted with appropriate drugs and other supportive measures as pain may make the child noncompliant to treatment and the parents to lose their trust in the treating personnel.*
- *Inadequate relief of pain has been found to lead onto post traumatic stress disorder, depression and stress even years after treatment.*
- *Clinicians often have misconceptions regarding the ability of children to perceive pain and to understand the nature and extent of pain completely.*
- *Pain is often undertreated in children due to fear of addiction to opioids and misconceptions regarding opioid usage and ignorance regarding pharmacodynamics and dosage of opioids leads to their inadequate and improper usage.*

- *Health professionals dealing with paediatric malignancies often lack knowledge regarding simple cognitive, behavioural and supportive techniques for pain management.*

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SUPPORTIVE AND PALLIATIVE CARE

METAPHORS AND BEYOND: A JOURNEY OF HEALING USING ART AND PLAY BASED INTERVENTIONS

***Chopra R**

Abstract: *Palliative care for children goes beyond just pain control and symptom management. It is a holistic model that also caters to the psychosocial aspects of the patient, their relationships, hopes, fears and wishes. Children seldom talk about their experiences, reactions, perceptions, feelings, wishes, wants or needs like adults do. Play is considered to be the natural language of a child. The symbolic language of play often reveals the child's inner world. This understanding can help the professionals to facilitate communication between parents and the dying child and thereby promote healing during the dying process.*

Keywords: *Childhood cancer, Play/art therapy, Healing, Psychosocial*

Hospitalisation can prove to be a very unpleasant experience for children and if the diagnosis happens to be that of a life threatening disease like cancer, it may even cause trauma. The inception of the treatment happens as soon as cancer is diagnosed leaving no scope for the children to prepare themselves for the gruelling interventions that are to follow. A series of side effects like recurrent pain and nausea, hair loss, weight gain/loss, body disfigurement, etc. follow once the treatment procedures of chemotherapy, radiation or surgery begin. The children have to undergo constant medical check-ups and hospitalisations disrupting normalcy and they are separated from their parents, siblings and friends in school eventually having a negative impact on their overall well being.¹⁻³ The unfriendly, unfamiliar environment at the hospital, sad and painful reality around may cause helplessness, fear, hopelessness, guilt, sleeplessness, decreased communication, sudden bouts of crying and in some children even depression.⁴⁻⁸ "Existence for them becomes a tortuous struggle between living and dying".⁹ As the disease

progresses, children often sense the gravity of the situation, anticipate death and fear separation from their loved ones which becomes the cause of anxiety, anger, despair or even depression in them.^{10,11} They are also observed to regress physically, psychologically, socially and emotionally.¹¹ This close encounter with terminal illness can make the child's happy world go topsy-turvy and eventually evoke confusing and conflicting emotions which needs an outlet for expression.

Unlike adults, children find it difficult to verbally articulate their feelings of traumatic unpleasant experiences through words. Their natural language to communicate is "play" which helps in developing a therapeutic alliance with the child in distress.⁴ Spontaneous play of a child with self and significant others is different from play therapy as the latter involves a professionally qualified therapist's relationship with the client in a specific environment. The British Association of play therapists define play therapy as "the dynamic process between child and play therapist in which the child explores at his or her own pace and with his or her own agenda those issues, past and current, conscious and unconscious, that are affecting the child's life in the present. The child's inner resources are enabled by the therapeutic alliance to bring about growth and change. Play therapy is child-centred, in which play is the primary medium and speech is the secondary medium".¹²

The play therapy room encompasses a variety of expressive and imaginative play materials like clay, art materials, dolls, puppets, sand play, board games, miniature objects, shapes, miniature animals, plastic media, symbols, etc. Stories, creative art and writing also are important tools in therapy. The environment plays a crucial role in therapy to be effective. Thus, the therapy space should be inviting and stimulating for the child to explore and experiment various possibilities and non-threatening and conducive to express one's thoughts and feelings freely.

A wide range of possibilities exist in using play as therapy. Therapy can be directive, non-directive or a collaboration keeping in mind the client needs. In directive therapy, the play therapist guides the child while in non-directive play therapy; the child is in control and decides what to do during the session within some gently and firmly

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set session boundaries.¹³ The choice of techniques by the therapist depends on various factors like the client needs, their age and most importantly the therapeutic objective. Some of the existing techniques of play therapy being used by professionals worldwide are: The Feeling Word Game, The Pick-Up-Sticks Game, Balloons of Anger, The Mad Game, Beat the clock, Worry Can, Spy and the sneak, Puppet-symbolic client (Kaduson & Schaefer, 1997), The slow motion game, Relaxation training- bubble breaths, Party Hats on monsters, Weights and Balloons, Broadcast news, The power animal technique (Kaduson & Schaefer, 2001), Color-Your-Life, (O'Connor, 1983), etc.¹⁴

Color-Your-Life technique was developed with the understanding that in order to cope with one's difficult emotions, there needs to be an awareness of various affective states. Children need to learn to identify their emotions in their day to day functioning and also express them verbally. With this purpose in mind, O'Connor used a drawing tool in which first the colours were paired with different feelings to increase awareness of various emotions and then the child was encouraged to draw all feelings they have experienced throughout life. This therapeutic technique provides a concrete way for the child to understand various affective states in a non threatening manner. This technique can be used at various phases of therapy to gauge the changes that have happened.¹⁴

O'Connor (1983) in his work of using this technique with a variety of children formulated some clinical impressions about this technique. Children who may be emotionally constricted or having poor self esteem may be using fewer colours and leaving large amount of white space on the page. The use of scribbles may indicate impulsivity and the portrayal of emotions in a stylistic, representational form as against using block patterns, etc. suggest ongoing emotional concerns with the child.¹⁵

In play therapy, art is just one of the many tools and techniques used. However, art as therapy is also being practiced as an independent intervention. Art therapy is based on the premise that visual art making is an important aspect of the human learning process as the psychological growth is enhanced with increased creativity¹⁶ and the creative processes involved in the making of art is healing and life enhancing.¹⁷ Visual arts involve making use of artistic tools like colours (paints, inks, pastel chalk, oil pastels, wax crayons, marker pens, colour pencils, charcoal, etc), clay, different kinds of paper, cards, envelopes, magazines, feathers, sequins, beads, wooden boxes/ frames, etc. to create an experience that is not only artistically pleasing but also marks a journey of self discovery for the client that triggers emotional catharsis.¹⁶⁻¹⁸ "Thus, art acts as a

'container' for difficult emotions".¹⁶ The tactile quality of art materials has a calming effect, enabling relaxation. Research also supports that art therapy helps in relieving anxiety and other emotional symptoms.¹⁷ Images speak through patterns, forms, colours, placements, size, positions, kind of material used, intuition; hence becoming a vehicle of expression for the child. Changes occur during the process of physical involvement with the materials, through the making of significant art object, through sublimation of feelings into the images and through communication with the therapist via the art object.¹⁶ There can be times during therapy when there is hardly any interaction between the therapist and the client. This is described by Winnicott (1958) as the "experience of being alone while someone else is present", which is an essential component for emotional development.¹⁹ Some of the visual art techniques being used by art therapists are: body maps, squiggle game, scribble chase, Mandala, etc.

Mandala is a Sanskrit word that means circle or centre and is considered a sacred geometric shape having no beginning or end.²⁰ It symbolises harmony, unity, wholeness and healing. The circle is filled with feelings and emotions that the child wishes to express at the moment in the form of colours, patterns and symbols. Once the bottled up emotions are expressed, healing begins. Drawing a Mandala does not have instantaneous reduction in anxiety or troubling emotions, but as per researches, drawing in a circular format does have a calming effect in the body in terms of heart rate and body temperature.²¹ Creating a Mandala is often a reflective process that enables a connection to one's innate healing abilities, which leads to personal growth and spiritual transformation.²²

Squiggle game is a technique developed by Winnicott (1971) in which the therapist involves the child in creating scribbles (squiggles) and then making drawings out of them and eventually making stories with the images created. This technique has its roots in the psychoanalytic approach as the client projects ones thoughts and feelings through art. It is also used to establish rapport with the child and communicate through drawing.

Having discussed about play and art therapy, it is now clear as to why children find it easier to express their unpleasant feelings and fantasies through the safe medium of play and expressive art materials than talking them out with the therapist.²³ "Unconditional positive regard and acceptance encourages the child to feel safe enough to be able to explore their inner selves without censorship. In this environment children are able to try out different roles, work through conflicting emotions and thoughts and try to figure out what the world is like".¹³ This helps them



Fig.1. Drawing by an 11 year old girl suffering from terminal cancer

overcome their fear and anxiety and facilitates healing as they distance themselves from the traumatic experience.²⁴ What to extract from the play and when is entirely the child's prerogative. Sometimes the play may just be a source of entertainment while at times a deeper exploration of the play space may also happen and children might sometimes draw connections with their real life.²⁵ Thus the therapeutic process of play enables the shift from the child's inner world to the outer reality.²⁶

The symbolic language of play and artistic creations often enables the therapist to peep into the child's world, their experiences, feelings and reactions. They also are able to gauge the child's true self perception and gain insights into their needs.^{27,28} Thus, every act of the child, the objects, symbols, colours used hold significance in therapy and must not be overlooked by the therapist. However, the interpretation of art work is directed by the client and not the therapist as the child is allowed to lead the process, giving them the choice of rejecting or correcting the art therapist's interpretations.¹⁹ So, the therapist does not rely on assumptions or perceptions alone but also brings forth and makes the child aware of any discrepancies and conflicting emotions that come across in the child's play or art and verbal and non-verbal expression.

"When a child is physically ill, body image is altered, self-esteem becomes lessened and the effects contribute toward a heightened sense of powerlessness".⁹ Children who have suffered physical changes due to the disease exhibit their reactions in their artwork by omissions,

exaggerations, making additional body parts, monstrous images which indicate conflict concerning body image.^{29,30} Feelings of inadequacy might sometimes be revealed in drawings in symbols like the absence of self.²⁹ Similarly, feelings of loneliness, isolation and anxiety may often be understood through images of body parts being omitted, the pressure of the line strokes, excessive shading, colors used, etc.⁹

To explore this further, I would like to share a drawing (Fig 1) made by an 11 year old girl suffering from terminal cancer. Due to the disease and its treatments, she had gained weight and developed scars all over her body. During the play session, she kept referring to her past when she looked pretty and everyone complemented her. Her focus during her interaction with the doctors was also solely on finding ways to get rid of the scars on her face.

This somatic preoccupation is also evident in the patient's drawing of the female figure with the red scars being prominently visible. This also suggests body image issues resulting in an impaired self esteem of the patient. The female figure looks like the patient's self portrait with the detailing in its features and clothing clearly indicating her preoccupation with self and a need for self display to gain social acceptance and acclaim. Her feelings of inadequacy and her need for physical power is also depicted in her drawing as she seeks affection, nurturance and warmth from others.

It is important to note here that the patient's vision about her creation was of utmost significance in guiding

me to understand the symbolism of her creation. My interaction with the patient along with my observations further helped me in interpreting her drawing. This also means that the therapist cannot completely rely on the standard interpretations of symbols in drawings as it is the meaning that each individual adds to one's creation that makes it stand apart from the rest.

Conclusion

Children suffering from cancer have a heightened need to express themselves and work through their day to day challenges. Play and art therapy makes this possible as the child is able to form a relationship with the provider and through this relationship they are able to develop trust, improve self-esteem and self efficacy. The therapist enables self awareness and acceptance for the clients elevating their confidence and the autonomy they acquire during play sessions is of utmost importance to them especially as they feel helpless in the hands of uncontrollable circumstances.³¹ Children eventually learn effective problem solving skills and master a situation by displacing their emotions onto an expressive art activity.²³ The difficult circumstances that a sick child is exposed to make them mature for their age; their childhood is often lost in the gruelling treatment procedures. However, one must not forget that he/she is still a child and can be helped to play as it is still their largest vocabulary.

Points to Remember

- *Only trained professionals should initiate play and arts based therapy with children.*
- *The act of simply having toys available for your clients or involving them in drawing does not mean that they are receiving (or that you are practicing) play therapy or art therapy.*
- *Change does not happen overnight. The process of therapy is more important than the outcome.*
- *There is no prescribed list of the materials/toys kept in the play therapy room. The choice of play materials has to be carefully done as some clients might be averse to some specific items.*
- *Even the sickest child can be helped to play.*

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CLIPPINGS

Continuous support for women during childbirth

Historically, women have been attended and supported by other women during labour. However, in hospitals worldwide, continuous support during labour has become the exception rather than the routine.

Primary objective was to assess the effects of continuous, one-to-one intrapartum support compared with usual care and secondary objective was to determine whether the effects of continuous support are influenced by: (1) routine practices and policies; (2) the provider's relationship to the hospital and to the woman; and (3) timing of onset.

Continuous support during labour had clinically meaningful benefits for women and infants and no known harm. All women should have support throughout labour and birth.

Hodnett ED, Gates S, Hofmeyr GJ, Sakala C. Continuous support for women during childbirth. Cochrane Database of Systematic Reviews 2012, Issue 10. Art. No.: CD003766. DOI: 10.1002/14651858.CD003766.pub4. Published Online: October 17, 2012.

Methods to improve healthcare worker hand hygiene to decrease infection in hospitals

Health care-associated infection is a major cause of morbidity and mortality. Hand hygiene is regarded as an effective preventive measure.

Objective was to update the review done in 2007, to assess the short and longer-term success of strategies to improve hand hygiene compliance and to determine whether a sustained increase in hand hygiene compliance can reduce rates of health care-associated infection.

The quality of intervention studies intended to increase hand hygiene compliance remains disappointing. Although multifaceted campaigns with social marketing or staff involvement appear to have an effect, there is insufficient evidence to draw a firm conclusion. There remains an urgent need to undertake methodologically robust research to explore the effectiveness of soundly designed and implemented interventions to increase hand hygiene compliance.

Gould DJ, Moralejo D, Drey N, Chudleigh JH. Interventions to improve hand hygiene compliance in patient care. Cochrane Database of Systematic Reviews 2010, Issue 9. Art. No.: CD005186. DOI: 10.1002/14651858.CD005186.pub3. Published Online: August 10, 2011

SUPPORTIVE AND PALLIATIVE CARE

NUTRITION IN PALLIATIVE CARE

* **Anjali Nair**

Abstract: *Nutritional support forms one of the essential adjuncts along with other measures of palliative care. To plan for proper nutrition of patients on palliative care, we need to assess degree of nutritional deficiency, type, quality of nutrients and decide about the mode of administration. The nutritive values of food items, various routes of feeding, their indications, advantages, etc are discussed in this article.*

Keywords: *Nutrition, Palliative care, Children*

Palliative care is a program of active compassionate care devised for terminally ill children, primarily directed towards improving the quality of life for them.

A multidisciplinary team that provides sensitive and skilled care to meet the physical, psychosocial and spiritual needs of both the patient and the family, involves physician, nurses, physiotherapist, psychologist, social worker, dietician and several other volunteers.

Good and adequate nutrition is the basic component for healthy living and more so during illness. Maintenance of nutrition and correction of nutritional deficits if any are major concerns in terminally ill patients especially when the illness is a malignancy and correction of nutrition is a major problem in terminally ill patients particularly when they are suffering from advanced malignant disease. Surgery, radiotherapy and chemotherapy can worsen these problems and need for improved nutritional intake is necessary for physical and mental health and proper dietary guidance is an integral part of patient care.

Causes of deficient nutrition can be many like anorexia, nausea and vomiting, mechanical obstruction to food, excessive loss of nutrients, cancer cachexia, adverse effects of the treatment taken, etc.

To help with proper nutrition for patients on palliative care, one needs to pay attention to -

- Assess the degree of nutritional deficiency, the type

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and quality of nutrients needed and the route and method of administration. Also one should note the effects and side effects of nutritional adjuncts for further continuation. Social and psychological needs of the patient and relatives are also to be taken into consideration.

Routes of feeding

Any one or more of the following routes can be chosen to administer the calculated quantity of nutrition to the patient.

- Oral route
- Enteral route or tube feeding.
- Parenteral route : Central / Peripheral vein.

Oral route is the best, simplest, cheapest and most physiological route to maintain the nutrition. Psychological factors like anxiety, depression and the attitude of care givers and relatives also contribute to the acceptance but the problems can be overcome, once the patient's appetite improves and it can be achieved by persuasion, attractive presentation of food, pleasant environment, correct temperature and aroma of food, small frequent feeds, taste tolerance and nutritional adequacy.

Enteral feeding or tube feeding is employed when the gut continues to have preserved mucosal digestion, pancreatic secretion and gall bladder emptying. Thus preserving mucosal integrity and reducing translocation of gut bacteria into circulation has lesser complications, infections and organ malfunction. It is cheaper than lesser parenteral nutrition.

There are three main methods of enteral feeding-

- Ryle's tube or nasogastric / nasoduodenal tube feeding: It is for short term-(<2 weeks), when there is a risk of aspiration. Ryles tube feeding is easy to start, simple to maintain and can be discontinued without any problem. Liquids can be administered.
- Gastrostomy feeding: Can be started easily and thick liquids can be administered.
- Percutaneous endoscopic gastrostomy (PEG) / jejunostomy are generally used for long term (>2 weeks).

Table 1. Recipes used as high calorie, high protein foods

Liquid diet	Soft diet	Full diet
Egg flip Milk shakes Plain milk Buttermilk Lassi Veg. thick soup Non-veg soups Cream soups Vegetable soups Coconut water Kheers Thick kanjis Dal water	All foods in liquid diet Porridges, kheer Khichadi, soft rice+ dal Mashed vegetables Boiled/scrambled/poached eggs. Lean meats Fish [pomfret] Chicken [without skin] Soft custards Puddings Plain gelatins Ice-creams	Everything accepted but restriction on spices, oil and fats.

It is therefore essential to think of some home made formulations of improved digestibility and texture which can be of better nutritional support than blenderized or elemental diets.

Therefore great emphasis is laid on malted cereals and legumes, which possess improved bio-availability of nutrients and at the same time provide nutrient dense and low bulk or less viscous diet which can easily be prepared from the available food materials (Table I).

Enteral Nutrition- With appropriate monitoring, complications associated with enteral feeding may be minimized or prevented. Enteral feeding complications can be categorized as-

- Mechanical: Tube migration, tube clogging, aspiration, gastroesophageal reflux.
- Infectious: Infective diarrhea, aspiration pneumonia.
- Metabolic: Electrolyte disturbance, micronutrient deficiency.
- Gastrointestinal: Nausea, vomiting, diarrhea, bloating and cramps, constipation.

Indications for enteral nutrition are: a) upper GI impediment, b) combined modality therapy, c) anorexia, d) psychological inability, e) inability to take large volume of oral feeds.

A registered dietitian will plan and prescribe ideal quantity and quality of food which can be prepared at home with the help of liquidiser, which can give all essential elements of nutrition without any intolerance and side effects.

Cancer, a devastating disease requires nutrition as its basic principle of palliative care in treatment of

terminally ill. Also the co-operation of relatives, with good and pleasant environment, consideration of cost and liking of patients, goes a long way in giving psychosocial support.

Guidelines

- Bed ridden children with enteral feeding tube: Different liquid diet recipes but strained can be incorporated in the enteral feeds.
- Very low appetite with mild dysphagia (head and neck): Start on calories dense liquid, (home based) feeds at small regular intervals, followed by full soft, semisoft bland feeds.
- Semi-conscious: Either full RT, NJT or NGT feeds are advised.

Points to Remember

- *Dietetic management is an essential part of palliative care.*
- *Among various routes of feeding oral route is the simplest, cheapest and most physiological one.*
- *Factors apart from illness to be considered to influence nutrition are preference of children, parents and relatives and pleasant environment.*

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SUPPORTIVE AND PALLIATIVE CARE

DEVELOPMENTALLY APPROPRIATE COUNSELING NEEDS IN PEDIATRIC PALLIATIVE CARE

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Abstract: *Aim and objectives: This paper aims to discuss and highlight the need for developmentally and age appropriate counseling for children and adolescents. Case studies are used as a qualitative tool to understand issues that need age appropriate counseling intervention for children facing advanced life threatening illness. Key issues regarding comprehension of their condition and coping devices to handle the resultant emotions are presented. The need for age appropriate developmental counseling is highlighted. Case studies are used to illustrate the child's understanding of his/her condition and the counselor's understanding of the latter.*

Keywords: *Cancer, Palliative care, Developmentally appropriate counseling.*

Cancer is emerging as a major cause of childhood deaths in Asia, Central and South America, Northwest Africa and the Middle East, where fewer children are now dying from preventable infectious diseases. Childhood cancer is generally not a public health priority in most developing countries. With the burden of HIV/AIDS, malaria and other infectious diseases - even the lack of clean drinking water - treatment for cancer is often regarded as unaffordable. In developing countries, many children who have cancer are never diagnosed, are diagnosed too late, or are diagnosed where treatment is limited or not available and hence incidence and mortality trends for childhood cancers are much more difficult to analyze due to inadequate reporting and competing causes of death. Childhood cancer usually refers to all cancers occurring in children before fifteen years of age.

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Although childhood cancers are rare, they are a leading cause of childhood death in developed countries.¹

A diagnosis of life threatening illness at any stage of child's life is a crisis for the patient and families in any country. The world of children and adolescents with chronic and life threatening illness is different from that of physically healthy peers.² Because the causes of illness and death for children are so different from those of adults, pediatric palliative care goals and treatments are also different. Parents and doctors must work closely together to make decisions that best benefit the patient while holding true to the values of the family. Pediatric palliative care focuses on the whole family since it is difficult to accept the mortality of children and parents may struggle emotionally and find themselves unable to cope. There may be siblings or other young children in the home who may face the impact of the child's illness in some way. The palliative care team looks for signs of stress within the family unit and provides interventions as necessary.

It is the right of child to have an access to compassionate care which offers relief from pain and addresses issues and concerns of patient and families through pediatric palliative care. In advanced phase of illness in children, the need for age appropriate counseling is vital and needs to be disseminated with sensitivity and the understanding that each child is unique and that human behavior is complex.

Fortunately today children and adolescents in India are extended the opportunity to exert the right to dignified treatment and care even in the advanced phase of illness. The Indian government plans to concentrate on cancer care in the next five years as announced by Health Minister in the Central Government on the occasion of the 27th National Cancer Registry meeting, where he also declared cancer as one of the non communicable diseases leading to increase in mortality and reiterated that as per National Cancer Registry Program of the Indian Council of Medical Research, the number of cancer cases is gradually increasing in the country.

The rising trend in cancer incidence and high mortality rate has caused concerns and posed challenges amongst the professional care givers as the patient population dealt

with is aware of their rights and informed about disease and its outcome. Hence, there is now an even higher need to understand coping mechanisms of patients as well as family members and to develop appropriate techniques while counseling both, in order to empower and equip them with required skills. This article thus aims to discuss and highlight key concerns of professionals while working with children and adolescents faced with advanced life threatening illness, as they seek intervention. The primary focus of this paper is to link the developmental age and stage of the child or adolescent and to select appropriate counseling approaches for them, to maximize benefit.

Pediatric palliative care of a premier cancer care hospital in india

The pediatric palliative care clinic of a premier cancer care hospital in india caters to 200 pediatric patients annually from age 0 to 18 years. The clinic aims to offer children and adolescents a platform to express their emotions without fear, guilt and shame, over and above offering medical intervention for their condition. The team has acquired training in handling current issues of concern and needs of the child or adolescent, carefully monitoring body language, developing insights through observation and of course completing a thorough clinical assessment. The focus is on recognizing the special needs of children with advanced cancers and making an attempt to offer developmentally appropriate care. Of great importance is their continuous availability to patients and families while the latter battle with crisis and face varied treatment dilemmas.

Patient and families referred to the pediatric palliative care clinic belong to diverse cultures and hail from different parts of the country. They present with a wide variety of linguistic, educational, cultural and socio-economic backgrounds, unique needs and different levels of understanding of their disease condition. On the other hand, they are also bombarded with global advances in media and the internet, which provide information about multiple treatment regimes, alternate therapies and engenders questions in the mind of the patient. This admixture poses an additional challenge for the multi professional team. Families irrespective of class, creed and religion come with a common goal of treating and curing their children.

The process: Pediatric patients with advanced cancer are referred to pediatric palliative care clinic after a combined decision of consultants through disease management groups (DMG). The treatment decision taken in the clinical meet is conveyed to parents by trained volunteers and counselors. The families are explained that the patient would now be taken through a transition to a new department viz. palliative

care, due to disease progression. The role of palliation is explained, the aim being management of symptoms and not disease cure, since the latter is no longer feasible in their case. Families have the choice to consult the parent unit to settle treatment queries and doubts if any. The palliative care clinic consultants mediate for patients and families to coordinate with parent unit if families have doubts and questions related to earlier treatment offered. The primary goal is to help the family come to terms with the finality of the decision by acknowledging their concerns, accepting them and then offering palliative treatment. This in turn leads to building up of a relationship, rapport and trust with patient and families, leading to their empowerment and improved compliance.

It is necessary to address the specific issue of choice to approach child and adolescent patients keeping in mind that they need developmentally appropriate practices and this is highlighted with a case study approach.

Methodology

Five case studies are identified, each at a different stage of development, in order to highlight issues and concerns. The case studies are selected with a view to depict developmentally age appropriate needs in children and adolescents while they suffer with life threatening illness like cancer. Case studies are a powerful qualitative method and help explore details of a phenomenon cited in context.³ Table.I helps provide a general guideline for the developmentally appropriate sequence along which cognitions and emotional responses develop in children and adolescents, in relation to death and dying.⁴

Case Study 1

A two year old girl child, a case of nephroblastoma treated adequately but did not respond to treatment. Patient was accompanied by grandfather and mother. The child though ill was active and playful. In the process of counseling the grandfather enquired about life span. The mother was a silent spectator and although dealing with her own in grief, had come to terms with it. The gender and seniority issues coming into play here, granting the grandfather more authority were clearly visible: at fourth visit he requested and mentioned that god should take the child away. He said they had no grudges against anybody but would wish the child a painless ending. The challenges in this case were minimal as family had already come to terms with reality of death. They only expected a less painful ending which we need to acknowledge as it is a natural response when little toddler's life is challenged.

In the above case the interventions were primarily

Table.I Developmental understanding of death

Age	Developmental Understanding	Death Concept	Examples & Explanations of Psychological Impact
1-3 years old	Death often seen as continuous with life. Death and Life are like Awake and Asleep	Does not understand irreversibility or permanence	The dead person is sleeping but will wake up
4-5years	Death is often seen as temporary and reversible; may also see death as a punishment	Death does not fully comprehend the concept of irreversibility or permanence; Believes in magical thinking or that you wish someone dead	Once you are dead are you always dead? "How long do you stay dead? I have been dead so now I have to die A dying child may struggle with guilt because he/she is dying.
6-9 years old	Understands death is permanent. Begins to realize people he/she knows will die and that dying means living functions will stop	Understands irreversibility and begins to understand finality or non functionality: heart stops you do not breathe, etc.	"Will dying hurt?" Is dying scary? What do you do when you are dead? A dying child may continue to experience guilt and shame for dying
10-13 years old	Understands death is permanent and that living functions cease. Begins to understand death is universal	Understands irreversibility, non functionality, and begins to understand universality.	Universality requires understanding three sub concepts: 1)Death is all inclusive, it occurs with all living things, 2)It is inevitable- eventually every living thing will die, and 3) It is unpredictable. The timing of death is not always certain. It is an unpredictable outcome in living things. "I am worried my mom will break down."
14-18 years old	Thinking becomes more abstract. One can objectively examine death. An adult understanding of death develops However, death may be viewed as an enemy that can be fought against Therefore dying may be viewed as a failure.	Understands irreversibility, non functionality universality and causality	Causality requires an abstract and realistic understanding of internal and external events that cause death. Events that cause death may be accidental or intentional, but often they are beyond the control of the dying individual. "I can't believe I am dying...Why this is happening to me"

acknowledging and accepting the family concern as valid. Counseling and reassurance that the best possible symptom management would be offered and efforts to keep pain in control would be undertaken constitute the primary counseling inputs. At the same time, the family needs to be told that despite great care unforeseen occurrence of emergency may always occur and hence false hope is not given. Here the child was not yet at a developmental stage to understand death and hence was stable. It must be noted here that not all children would understand what happens to them. Parental concerns are vital and must be addressed, eg. the child may be an only child and hence more precious, or parental age may ensure that it is now impossible to plan for another baby. Parental anxiety may further mount due to the child's inability to verbalize pain. The child may be irritable or tearful and families do not understand how to deal with this.

Case Study 2

A six year old girl with neuroblastoma was on treatment over a year with surgery and chemotherapy. Her disease relapsed and she was referred to pediatric palliative care services for symptomatic treatment. She was brought for treatment to Mumbai as the place from where she temporarily migrated for treatment did not offer specialized oncology care.

While she suffered in advanced phase of illness, she expressed anger against god and pleaded god to either take away her life or take away her pain. She often questioned her mother "Why does god do this to me? "Why does he not relieve me? She could feel something grossly wrong with her body and hence subtly communicated her awareness about prognosis while she lay in her bed immobile. Sibling envy and rivalry was noted as parents informed the counselor about her tantrums and fights with siblings. She desired to attend school and wished to be normal like all other kids of her age in school. Confined to her bed, she envied all those who could move about and play. She enjoyed total attention of her parents; the healthy siblings had accepted the fact that she needed attention due to her illness which they found frightening. They were confused because they had mixed feelings, resenting the attention given to her, but guilty about this resentment since they could see she was ill and required it.

The family was unaware about how to manage the situation. They were stressed and had no answers to her questions. The choked and anticipated grief appeared in the form of tears. The only answer they offered to the girl was the hope that her treatment was on and she would feel better. The parents experienced strong feeling of guilt as

they felt they had let the child down, failed to keep her safe or make her better despite all efforts. The counselor needed to explain to them that she was struggling to understand her own condition. Children, with their age appropriate understanding, often attempt to communicate awareness of what is happening to them. Not all have the vocabulary to express emotions. Therapeutic intervention in the form of play, art, craft, drawing, pictures, stories and narrations of illness trajectory can be helpful to reach out to them. The same can then be conveyed to parents, as an alternative language of communication with the child. An unconditional acceptance, reassuring touch or pat, or even silence helps professional caregivers win over these wounded warriors who need anchors to guide them. If the same approaches can be taught to parents, the gains are twofold. The parents and child both have a sense of satisfaction at having communicated their innermost feelings to one another. And both also develop a new sense of peace since they are no longer struggling with barriers between them. The parents come to realize that many of the child's questions may not really have any answers and all they have is themselves, their reassuring presence, to give to the child, in order to tide over this painful period and save up memories for themselves.

Case Study 3

A ten year old boy, a case of "Primitive Neuroectodermal tumour (PNET)" treated with multi modal treatment relapsed and was advised palliative care. He was regularly followed up with palliative care clinic and was very happy with treatment. However his body language and expressions spoke of his inner turmoil and his concerns about and awareness of prognosis. One day he managed to enter pediatric palliative care clinic out of turn while his mother was away. On entering the consulting room of doctors and counselor he placed his hospital chart in front of the consultant with the recent report of PET scan which indicated increased size of older nodes and uptake indicating development of new nodes. Calmly the pediatrician with a warm glance at him enquired where his care giver was; he replied that his mother is coming as she has gone to complete some formality. He then requested the consultant to explain to him his PET CT report. The pediatrician asked him what he understood from the reports and he replied that it seems that the old nodes have grown in size and new nodes have appeared. The pediatrician nodded in acceptance and questioned again what this meant to him. The boy replied that the disease seems to have grown. The consultant was about to continue the conversation, but taking advantage of an unexpected interruption midway of the conversation the boy shut his file and said okay this is all and left the room.

He revisited the consultant on his actual turn with his mother. He was calm and quiet, in the process of counseling he mentioned that he wished to stay with his grandparents in the farm. He shared that he did not like crowds and people enquiring about his illness and also did not like school and friends.

His case depicts how children wish to be partners in disclosure and care. He exerted his autonomy of choice of place to stay and live. His awareness that the elders will not object to his staying in the farm and that his grandparents loved him unconditionally irrespective of illness and its impact, perhaps led to this decision. His choice of not going to school to avoid friends and related illness discussion was respected. He opted to go back to his village as he understood treatment limitations and had come to terms that disease is progressing. He may not have understood severity of terminal illness and had not verbalized it. He had not expressed concerns of terminal phase probably because of lack of information or maybe he did not wish to know. The issue of diagnostic or prognostic disclosure to children and partnering in treatment decision is debated and researched all over the world. Findings indicate that some parents do not wish to inform children and involve them in treatment decisions, despite the known fact that children are aware of treatment outcome. Parents in cases like these find it very difficult to grant to a child the autonomy of decision making. They are still struggling with their own conflict of hope versus despair and continue to blindly hope for survival in spite of medical prognosis to the contrary.

Case Study 4

A 17 year old girl treated for osteosarcoma of right femur was accompanied by her father. Her mother is no more, her two younger siblings stayed alone in deep interiors of the state while she was treated at the cancer care hospital.

She presented as satisfied with happy facial expressions despite her illness. Her goal was expressed as 'to get cured, complete education and become a teacher to offer quality education to village children and care of the younger siblings in the absence of their mother'. While on active chemotherapy post surgery, she relapsed and developed lung metastasis. She was advised symptomatic care. The hospital in those days did not offer palliative care service to children. It was challenging communicating with her, as her father insisted that she be informed about her prognosis by the professionals, since he did not have the courage to face the situation. Distressed adults may have difficulty in talking to children or adolescents. If the news is not good they will want to protect them from hearing what they really do not want to tell them. Adults often have

fears and concerns about who should tell the child, when, what the child needs to know and how to say it. Consideration must be given to the child's possible reactions and how adults will respond to support the child.⁴

The girl predicted the unexpected truth of her limited life from her father's tearful expressions, she went up to her counselor and said "I am not worried about my health and limited life span which I am prepared but am feeling bad that my siblings will be losing their mother once again." She cried saying that her father did not remarry as they were too small and felt that the step mother would not take good care of the children. She became mother of her younger siblings at tender age of eleven and life moved on. Now she felt, while they had begun to cope with the loss of their mother, cancer struck her, a surrogate parent. She felt bad for her younger siblings who would be orphaned once again. The case highlights at the same time, the innate resilience of children and adolescents, more so in India where fatalistic attitudes are often engendered by the culture and spiritual outlook. In India many rural children grow up and mature to shoulder familial burden of survival.

This case study further highlights the need of pediatric palliative care services and need to address adolescent issues as they face dual crisis of transition and advanced disease. They no longer will get the opportunity to enjoy like normal adults. In the advanced phase of illness the counselors need to understand multiple issues such as those of identity, unattained goals, resilience and age appropriate understanding of life, becoming an untimely adult, break in education, isolation from peers and families. The importance of appearance, loss of hair, beauty, independence, etc. which are of paramount relevance to the adolescent are in cases such as these, pushed aside to make place for responsibility toward the family and social circumstances. These choices are those made due to inevitable social circumstances, poverty and lack of services.

To help adolescents cope effectively with their experiences of cancer, health professional's need to develop insight into the challenges that adolescents endure and an understanding of what resources each adolescent needs. Each adolescent differs in their response to demand and opportunities of growing up. Biological, social, emotional and intellectual growth may occur at different times and rates throughout adolescence. This may affect the diagnosis and prognosis greatly posing, further challenges.⁵

Case Study 5

A 19 year old adolescent boy treated for ALL relapsed

while on treatment was referred to palliative care and was accompanied by his mother a widow and grandmother and an adolescent sister. In his first counseling session on the day of referral he was not aware of his prognosis. He came to palliative care following referral by his doctor. His mother was aware of the prognosis, grieved it and informed the counselor that he is a grown child and understands everything. He has been managing himself all through treatment process. He was pursuing his junior college education while his mother was engaged as a casual laborer in the village. The counselor was thus given an impression that the youngster would be resilient to poor prognostic information. However, in the process of counseling it was observed that the boy knew about relapse but could not relate it to bad prognosis and life threatening outcome. He was shocked to hear the news and cried but adjusted quickly. Issues of self harm to hasten death were discussed openly and he reassured the counselor that he did not wish to harm his mother emotionally any further by taking any such drastic steps which would make her unhappy. Counselors often avoid discussions of death by one's own hand and self harm, worrying about "planting ideas" in the child's head. However, one needs to pick up pointers about whether such thoughts are already present and hence addressing them freely and frankly would perhaps be much better. In this case, the patient went back to his native place, deteriorated very fast and passed away within a couple of weeks.

Discussion

Each of the above cases is a glimpse of the patient's expressions and reactions in advanced phase of illness. There are many such cases which demand the need to communicate and empower patients and families through counseling. We cannot counsel children the way we counsel adults. We need to engage them in a way that they talk freely about painful issues. Children at any stage of developmental cycle have an age appropriate understanding of what happens to them. A child two year old is concerned about the mother's facial expressions and tears. A little older child, above five years of age would feel separation anxiety; they learn to verbalize as they grow up, but many are unable to verbalize due to poor communication and social skills and yet they are curious about the surroundings. The hospital environment may create fear and insecurity, which would need to be addressed.

Counselors while counseling the older adolescent patients attempt to touch upon very tender issues of self harm keeping in mind the age of patient and his/her understanding of prognosis and related issues that bother

them. The counselor's judgment and observational insights, the patient's mood and cooperation in responding to questions and initiative in counseling process are all very vital skills that help make counseling session a fruitful exercise. If patient's body language or questions hint towards a warning of ill intentions then care is taken to counsel the family members about drug management schedules, placing medicines away from patients, or keeping away sharp objects which can be misused during a bout of excruciating pain.

The need for age appropriate counseling is paramount, as each child is different and human behavior is complex. Some commonalities established through observation and development of psychological models is of great help in the counseling process. The child's developmental level should be assessed to identify ways to help the child understand and accept the implications of illness and possibility or inevitability of death. Hence, not just chronological age, but mental or intellectual age needs to be looked at. Child development is a continuum and a child with a chronic illness may regress developmentally or may have been developmentally a little slower to start with. A basic understanding of children's developmental stages regarding issues of health, illness, death as well as spirituality and religious beliefs is important before discussing psychosocial issues.

Parents may have justifiable concerns that discussing these issues will increase a child's fears, and that too must be laid to rest. They need to comprehend that giving a child developmentally appropriate opportunity to be knowledgeable about his or her condition does not necessarily heighten death anxiety. Allowing a child to discuss different aspects of an illness may decrease feelings of isolation, alienation and the sense that the illness is too terrible to discuss completely.

Many children seem to know when they are dying, a fact very apparent to researchers who with parents permission have talked to very ill children. Some of this research indicates that children even as young as two years old can feel fear of dying in the form of being separated from parents and other loved ones and these fears need to be allayed. Another study about how children talked about the visual and verbal images to describe death, led to the interesting finding that older children said they would want to know the age of the child they were trying to explain death to, so that they could use gentler images and not scare the younger one⁶. This is a very common desire shared by all patients and care givers: the wish to protect each other from the harm of disclosure or discussing death and dying.

Conclusion

“Nobody knows what tomorrow holds for the treatment of cancer. There are still many more unanswered questions than answers, but one day, perhaps still in the far future all the questions will be answered and all the mysteries will be solved. It is certain that today’s treatments are more effective than yesterday’s and that tomorrow’s will be even better.”⁷

Points to Remember

- *The role of palliation needs to be explained to parents. The aim is management of symptoms and not disease cure, since the latter is no longer feasible.*
- *The goal is of building up a relationship, rapport and trust with patient and families, leading to their empowerment and improved compliance.*
- *Unforeseen occurrence of emergency may be there and false hope is not given.*
- *For younger, less verbal children, therapeutic intervention in the form of play, art, craft, drawing, picture, stories and narrations of illness trajectory can be helpful and the same can be taught to parents.*
- *Older children may wish to be partners in disclosure and care and exert autonomy of choice of place to live.*

- *Innate resilience of children and adolescents is apparent, often more so in India where fatalistic attitudes are often engendered by the culture and spiritual outlook. In India many children grow up and mature to shoulder familial burden of survival.*

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CLIPPINGS

Early trophic feeding versus enteral fasting for very preterm or very low birth weight infants

The introduction of enteral feeds for very preterm (< 32 weeks) or very low birth weight (< 1500 grams) infants is often delayed due to concern that early introduction may not be tolerated and may increase the risk of necrotising enterocolitis. However, prolonged enteral fasting may diminish the functional adaptation of the immature gastrointestinal tract and extend the need for parenteral nutrition with its attendant infectious and metabolic risks. Trophic feeding, giving infants very small volumes of milk to promote intestinal maturation, may enhance feeding tolerance and decrease the time taken to reach full enteral feeding independently of parenteral nutrition.

Objectives of this review was to determine the effect of early trophic feeding versus enteral fasting on feed tolerance, growth and development, and the incidence of neonatal morbidity (including necrotising enterocolitis and invasive infection) and mortality in very preterm or VLBW infants.

The available trial data do not provide evidence of important beneficial or harmful effects of early trophic feeding for very preterm or very low birth weight infants. The applicability of these findings to extremely preterm, extremely low birth weight or growth restricted infants is limited. Further randomised controlled trials would be needed to determine how trophic feeding compared with enteral fasting affects important outcomes in this population.

Morgan J, Bombell S, McGuire W. Early trophic feeding versus enteral fasting for very preterm or very low birth weight infants. Cochrane Database of Systematic Reviews 2013, Issue 3. Art. No.: CD000504. DOI: 10.1002/14651858.CD000504.pub4 Assessed as up to date: December 27, 2012. Published Online: March 28, 2013.

SUPPORTIVE AND PALLIATIVE CARE

COMPLIMENTARY AND ALTERNATIVE THERAPY - YOGA IN PALLIATIVE CARE

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Abstract: *Yoga is an ancient Indian practice of healing mind, body and soul. Today, yoga has become one of the popular areas of research and is being effectively used as a component of Alternative and Complimentary Practice of palliative care for many ailments. What is unique about the practice of yoga is that it is not confined in its application and people from all age-group can benefit from it. In the recent years, with its wide application in the field of education and health, yoga has become an integral part of learning and practice in children's lives and recent research focuses on studying beneficial effects of yoga on children.*

Keywords: *Yoga, Children, Cancer, Alternative and Complimentary Medicine, Mindfulness Meditation.*

A diagnosis of life-threatening disease like cancer brings with it a lot of physical and psychological suffering for young children. An array of painful symptoms, distressing treatment procedures, considerable anxiety due to the uncertain nature of disease and constant emotional trauma become a part and parcel of their journey through Cancer. To make their journey less distressing and smooth and to bring about the optimum expected results of the treatment, healing the mind and spirit using "Integrative Therapies" along with treating their bodies become very essential. Yoga is one such effective therapy which is believed to heal mind and body.¹

Yoga, a Sanskrit word meaning "to unite-to integrate" is an Indian body of knowledge and an ancient practice for attaining mind-body balance, spiritual awakening and healing. Discipline of yoga has three pillars on which it rests for achieving inner sense of balance with positive effects for mind, body and soul that get visible over time. These three pillars are: Breathing Techniques (*Pranayama*), Postures (*Asanas*) and Meditation (*Dhyana*). The methodical

combination of practice of yoga asanas, pranayama and dhyana is said to evoke a feeling of peace and tranquillity within an individual. Practice of yoga facilitates relaxation, improves sleep and boosts positive feeling. Thus, it works as an effective stress management technique. Yoga also improves blood circulation and oxygenation in the body which helps maintain normal heart rate and blood pressure.²

Yoga has been practiced for over 5000 years and from the time of its origin till date, yoga has evolved as a healing practice and crossing cultural and regional boundaries, today more than 11 million Americans are enjoying the health benefits offered by yoga. Noticeable research is being done in India as well as abroad to study the physiological, biochemical and psychological effects of yoga and today its therapeutic powers are integrated with modern medicine to help patients suffering from heart ailments, mental disorders, skin ailments, and even life-threatening disease like Cancer.³ Though yoga has ample benefits, it can become a risky practice if practised without proper coaching and training. Thus it is suggested that yoga must be learnt and practised under supervision before one begins to practise it independently.⁴

About benefits that yoga offers, it is established that yoga can enhance muscle strength, tone and flexibility. It helps improve balance and posture. It aids breathing, promotes relaxation and improves lung capacity. It also helps in lowering stress and improves concentration and mood. Yoga is known to benefit the heart by lowering blood pressure and heart rate which aid better heart function. Research suggests that yoga helps in decreasing cholesterol and triglycerides and boosts immune system functioning.⁴

It is suggested that yoga has something to offer to everyone from across all age. Yoga can benefit every child across any age-group ranging from toddlerhood to teenage. Recent research is focusing on studying beneficial effects of yoga for children suffering from various problems, disorders and diseases. Encouraging anecdotal reports describe yoga as helpful tool for calming children, reducing obesity in children, mitigating behavioural problems, alleviating anger, aggression, anxiety, depression and panic attacks among children, and enhancing their imagination, concentration, and academic performance.⁵ Health problems, such as headaches, stomach aches, constipation,

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back pain, and colds or sinus problems, are reportedly improved with yoga practice as well as a decreased need for medication for children with attention deficit disorder.⁵ Literature suggests that there is limited but growing empiric support for positive health effects of yoga.⁵ Review of available literature on yoga for children acclaims that yoga has positive effects on cardio-vascular functioning, physical endurance and strength and behaviour.

Components of teaching yoga lessons to children include relaxing environment, soothing atmosphere, and appropriate duration of the sessions and activities in the session.⁶ Children can very well relate to the objects in the Nature and in their surroundings like animals, birds, flowers, trees, mountains, etc. thus, a yoga instructor can encourage children to relate to these objects and expand their imagination as well as teach some postures easily by helping them relate to familiar objects. Music, chants, prayers and meditation can be well integrated to aid relaxation of mind and enhance experience of peace and joy.⁶ While connecting with the nature, children are able to connect with their own selves and this helps in relaxation, better understanding of self and others too.

Yoga mats, pillows, stuffed animals can be used extensively in children's yoga classes. Another important thing is safety of children and thorough supervision. The temperature of the room or the yoga postures, exercises and breathing techniques should not prove strenuous for children and in conditions of discomfort or pain children should be allowed to discontinue the practice.⁶

Yoga is found beneficial for children in school setting as well in hospital settings. Thus, it is suggested that when it comes to providing palliative care and using alternative and complementary medicine for children, techniques like yoga should be introduced early in the course of children's illness and treatment, for it to be more effective and help children feel more empowered during the challenging phase of their sickness. St. Mary's Healthcare System for Children, Bayside, New York is one such institute which has remained open to using alternative healing methods for their pediatric patients. It offers classes in Tai Chi and Yoga to help children focus and develop their inner strength which enables them to get through painful phases of their illness.⁷

Another evidence to validate effectiveness of yoga is provided by OTs and PTs at the Children's Hospital Medical Center in Seattle, WA. They have developed a special yoga program which is very aptly called Bendy Kids that holds two classes in a week for pediatric oncology patients. Specialized instruction is aimed at improving flexibility, balance, respiratory function, endurance, sleep patterns and

improving general strength and vigour-a definite asset for children enduring cancer treatment regimens. (www.seattlechildren's.org).⁸

In recent times, "Mindfulness Meditation- a mind-body intervention" is studied and being used in pediatric clinical practice. Mindfulness Meditation is a technique which can be learned by all and when practiced in disciplined manner is found to aid stress reduction and foster better pain control.⁹

The Children's Hospital of New York- Presbyterian is another such hospital which offers complementary and alternative therapies to pediatric oncology patients. It offers yoga classes for children twice a week in outpatient clinic. yoga classes here emphasize on deep breathing, stretching exercises and meditation which is believed to foster relaxation and healing. Yoga also helps achieve flexibility, strength, balance and co-ordination and emotional stability. Dr. Kelly's research found that 85% of pediatric oncology patients at Columbia used complementary therapies and she also found that half of those families had not informed their oncologists of their use of supplements.¹

Dr. Kelly's findings suggest that there is a considerable need for research in establishing effectiveness of "Complementary and Alternative Medicines (CAM)", making health care professionals aware about its effectiveness and making them proactive in integrating CAM with main stream medical line of treatment. Also, parents of pediatric patients need to be educated about how to use CAM safely and effectively with their children and how to ensure support of medical professionals in using it.¹

Points to Remember

- *Yoga, an Indian body of knowledge is widely used and practiced around the globe to achieve mind-body balance, spiritual awakening and healing.*
- *The therapeutic powers of yoga are integrated with modern medicine today and it is suggested that it is very effective in providing comfort and in some cases cure, to the patients with various ailments and life-threatening illnesses.*
- *Yoga is beneficial for everyone be it children, young adults or senior citizens.*
- *Yoga is evolving as an adjunct to healing practice and there is great scope for research in this area especially to understand its effects among children and adolescents suffering from life-threatening illnesses.*

- *There are Children Cancer Hospitals in the US who have adopted yoga practice and offer it to their pediatric patients.*
- *“Mindfulness Meditation- a mind-body intervention” is studied and being used in pediatric clinical practice today.*

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CLIPPINGS

Rojas MA, Lozano JM, Rojas MX, Rodriguez VA, Rondon MA, Bastidas JA, et al . Prophylactic Probiotics to Prevent Death and Nosocomial Infection in Preterm Infants. Pediatrics Nov 2012.

Although *L reuteri* did not appear to decrease the rate of the composite outcome, the trends suggest a protective role consistent with what has been observed in the literature. Feeding intolerance and duration of hospitalization were decreased in premature infants <1500 g.

GENERAL ARTICLES

SYNCOPE IN CHILDREN***Chokhani Rajesh R**

Abstract: *Syncope is not uncommon in children. Since the list of causes is large, a sound clinical approach is necessary to effectively separate out syncope from its mimics, and then zero down on the probable diagnosis. While vasodepressor syncope is the commonest and benign, it is crucial to identify potentially life threatening cardiac syncope as well. This article attempts to outline a practical approach, which on one hand limits unnecessary investigations, while on the other, effectively screens for serious disorders.*

Keywords: *Syncope, Pediatric, Vasodepressor, Cardiac*

Health care providers are quite familiar with the scenario where a child is rushed to them because he / she suddenly collapsed in the bathroom or in school. Such a complaint could be a syncopal attack; syncope is not uncommon in children. The real challenge while dealing with such complaint is to differentiate the benign from the dangerous. And in doing so, one needs to strike a delicate balance - evaluate enough so as not to miss a potentially dangerous condition, and yet, avoid unnecessary investigations where, in all probability we are dealing with a benign condition. In clinical practice, this balancing act is not easy, but with a systematic approach and understanding of the subject, it can be done with reasonable confidence and accuracy.

Approach

While evaluating such a patient, the two main questions that one needs to try and answer are:

1. Is the presenting complaint really a syncopal attack or something else (seizure, breath holding spell, conversion reaction, vertigo) ?
2. If it is a syncopal attack, is it vasodepressor (vasovagal), cardiac or other ?

The most important tool that helps to answer these questions in a large majority of patients is a detailed history.¹ Almost always, by the time the child is brought to the doctor, there may not be any signs to go by. Similarly, investigations rarely help to narrow down the primary diagnosis. Thus, one cannot expect to differentiate a 'fit' from a 'faint' in a given patient by asking for both an EEG and an ECG, or for that matter, other investigations. Thus, investigations may have a role only after a provisional diagnosis has been made, in an attempt to confirm or rule out a specific suspected condition.

History taking

The first and foremost purpose of the history is to get the presenting complaint right. Parents/caretakers may use colloquial language and thereby describe what happens to be their own perception and analysis of the event, and possibly mislead the 'busy' physician. Therefore, it is best to ask them to describe what exactly happened. A detailed, second by second account of what exactly happened is crucial in making the correct diagnosis. More often than not, the parent/caretaker who brings the child to the doctor has not been at the scene of the event. If so, it is worthwhile to question the teacher/ friend/other child/adult who witnessed the event, either personally or even over phone, preferably as early as possible, so that the event is fresh in their memory. Further, one must never fail to question the child patients; once again, contrary to popular expectations, they can and do provide significant information. Finally, one has to almost 'cross examine' the witnesses/child patients, so that the final account that emerges is just about correct, without any misleading additions/subtractions.

No doubt there are limitations to such an exercise¹, but that alone should not put us off; because if an accurate detailed description can be obtained, it can be far more valuable than a bunch of expensive investigations.

Mimics - differentiating syncope from other events**Seizure**

The most important distinction that needs to be made is between a 'fit' and a 'faint'. A seizure may be preceded

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Table I. Seizure vs Syncope

Seizure	Syncope
Aura	Prodrome
Tonic/clonic movements occur before LOC	T/C movements occur after LOC
Bladder, bowel incontinence	No incontinence
Skin - warm, flushed and /or cyanotic	Skin – pallor, sweating
Eyes open	Eyes closed

by an aura, the tonic or tonic/clonic movements occur before the child loses consciousness, there maybe bladder or bowel incontinence and the skin may be warm, flushed or depict cyanosis². What is extremely important is that during a seizure the patient's eyes are open, as against in a syncope where the eyes are closed (Table I).

Anaphylaxis

A child can develop anaphylaxis following infections, which may be mistaken for syncope by the health care provider. A diagnosis of anaphylaxis should not be missed. Presence of angioneurotic edema, rashes, tachycardia and weakness of central or peripheral pulses will point to anaphylaxis.

Breath holding spell

A breath holding spell can be easily separated out by the classic history of an emotional trigger – pain, anger, or fright, following which the infant/child (age 6 m – 5 years) starts crying loudly, then holds the breath (clinically marked by the ‘muting’ of the cry - while the child's mouth is open, there is no audible cry), which is then followed by cyanosis/pallor, which can then be followed by a transient loss of consciousness and occasionally, even a few convulsive movements. Pallid attacks share many similarities but the child looks pale instead of cyanotic.

Conversion reaction (hysteria)

These episodes typically occur in the presence of an audience, are often prolonged and are not posture dependant. The child does not sustain any injury and intelligent questioning can demonstrate that the child ‘remembers’ the event/environment even during the phase of alleged loss of consciousness.

Vertigo

Vertigo can be confused with syncope only because

the narrators may use words which suggest the former, though an accurate description of events settles the issue. Vertigo is likely to be less sudden, more prolonged, the child will clutch onto something to steady himself/herself and thereby prevent a fall, and there will not be a clearly definable period of loss of consciousness.

Syncope

Syncope is defined as the sudden loss of consciousness and postural tone with spontaneous and complete recovery after a brief duration.¹

Theoretically, there is a big list of causes of syncope in children, which can be referred from any standard text. However, vasodepressor syncope (also known as vasovagal or neurocardiogenic syncope) is the most common (and also benign) cause of pediatric syncope. Cardiac abnormalities, though uncommon, are important because they can be life threatening. Besides these, rarer causes, where the presenting complaints are likely to be different, include neurological causes (seizure, migraine, hyperventilation syndrome), metabolic causes (dysselectrolytemia, hypoglycemia, endocrine disorders, drugs, toxins) and post tussive syncope.

Thus, for all practical purposes, one should learn to clinically separate out syncope from other similar looking events, and then go on to identify vasodepressor syncope, taking care not to miss out the occasional cardiac syncope.

Clinical features of vasodepressor syncope

What the child experiences.....

The child may experience any or all of the following: a warm, hot or cold sensation, lightheadedness, dizziness, nausea, hearing or visual change (loss of hearing, rushing noise, blurring of vision, darkness, tunnel vision, or double vision), an anticipated loss of consciousness (weakness and a feeling of going to the ground). After regaining consciousness, the child may feel weak, groggy and tired³.

What witnesses notice.....

The patient suddenly loses posture, may appear pale or ashen, may be sweating, and the skin may feel cold. There can be brief tonic, clonic movements, though there is no incontinence. Classically, the loss of consciousness lasts for just a few seconds, almost always less than a minute.³

The setting - Precipitants that contribute

Age and sex: It is more common in girls and teenagers

Position: prolonged upright posture, position change to more upright

Time of the day: It often occurs early in the morning, as soon as the child gets up from bed and goes to the washroom; the child may 'collapse' while brushing teeth or while micturating. It is also common in the morning school assembly or during physical education in the school playground in the harsh sun.

Last meal: There is often a history of a poor breakfast that particular morning.

Emotional triggers: Pain (including vaccination, particularly in teenagers/adolescents), fear, sight of blood, smells, anger.

Other factors: Anemia, relative dehydration, hunger, recent or present illness, physical exhaustion, crowded, poorly ventilated confines

Situational syncope: Some patients have stereotypical triggers such as voiding, instrumentation, post exercise, pain etc that lead to recurrent syncope.¹

Red flags that point to cardiac syncope

Young age: Vasodepressor syncope is rare below 6 years of age

Exercise induced syncope: A syncopal attack that occurs during exercise suggests a cardiac cause; such a syncopal attack is aptly also known as 'mid stride' syncope. This needs to be differentiated from a syncopal attack that may occur immediately after a bout of heavy exercise.

Palpitations/chest pain: The child may be able to describe a feeling of racing heartbeats or the caretakers may appreciate the same with their hand on the child's chest, thereby signifying a tachyarrhythmia. A complaint of chest pain may signify cardiac ischemia.

Family history: A family history of sudden death, SIDS, drowning, pacemaker placement, or recurrent syncope strongly suggests the need for extensive cardiac evaluation.

Past history: A history of congenital heart disease or cardiac surgery also necessitates cardiac evaluation.

Absence of a prodrome also points to an increased risk of serious heart disease and needs detailed evaluation^{2,3}.

Physical Examination and ECG

While a detailed physical examination is mandatory, specific attention should be paid to physical features associated with cardiac disease (like abnormal facies, Marfan habitus, deafness, ataxia) or neurological disease (like ash leaf spots, café au lait spots, cleft palate). The cardiovascular examination should focus on orthostatic blood pressures, auscultation in supine and standing positions and the cardiac rhythm.

Table II. Important ECG findings in pediatric syncope

Long QT syndrome	QTc > 0.45 msec
Wolff Parkinson White syndrome	Short PR interval, wide QRS complex, and a delta wave or positive slurring in the upstroke of the QRS complex
Hypertrophic cardiomyopathy	LAE and LVH, ST segment abnormalities, T wave inversions, Q waves and diminished or absent R waves in lateral leads
Brugada syndrome	ST elevation in anterior precordial leads, V1 & V2, type 1 coved

An ECG should be obtained in all pediatric syncope patients. Though the yield is low, it is inexpensive, non invasive and yet, highly sensitive. In combination with a detailed history and a physical examination, it has a sensitivity of 96% for picking up cardiac syncope. Besides confirming suspicions that may have been raised by the history or physical examination, it has the potential to provide clues to the presence of those cardiac disorders that may have led to the syncope but yet, may not have any physical findings at the time of evaluation.

In case the primary pediatrician is not confident of reading the ECG, it may be sensible to get it read by a specialist⁴. An incorrect interpretation of the ECG can lead to both errors of commission and omission.

Thus, if the history is typical of vasodepressor syncope and the physical examination and ECG are normal, the patient does not need further evaluation. On the other hand, if there are red flags that are identified in the history, and/or physical examination is abnormal and/or the ECG is abnormal, the patient needs a detailed cardiac evaluation.

Physiology of vasodepressor syncope

The physiologic disturbance that leads to syncope could be primarily hypotension, primarily bradycardia or a mixture of both. Over the years, it has been believed that the Bezold Jarisch phenomenon leads to these disturbances. Forceful contractions of an under filled left ventricle leads to excessive stimulation of the ventricular mechanoreceptors or C fibers, which in turn leads to paradoxical signals to central nervous system pathways. A sudden conversion from vasoconstriction to vasodilatation and bradycardia then follows. However, of late, it is increasingly felt that children

with syncope form a heterogeneous group and there could be more than one causative mechanism. Thus, aberrant autonomic regulation, endogenous vasodilators, disordered baroreflex function, paradoxical cerebral autoregulation, and low serum ferritin may all play a vital role.¹

Treatment of vasodepressor syncope

Non pharmacological treatment remains the mainstay of treatment of vasodepressor syncope. The child and the family should be educated and reassured. Adequate hydration should be encouraged and is found to be 90% effective. Enhancing dietary salt intake and avoiding caffeine may help.⁵ Potential triggers should be identified and avoided if possible. Children should learn to recognize their prodromal symptoms and promptly sit down or lie down for a few minutes till symptoms resolve. Post vaccination syncope in adolescents can be avoided by vaccinating in supine position or insisting that they lie down for few minutes immediately after vaccination before they leave the clinic.

Pharmacotherapy forms the second line in the management of vasodepressor syncope. Midodrine, β blockers, fludrocortisone, specific serotonin reuptake inhibitors appear to be useful treatment options. While most of these agents are presumed to be effective, randomized controlled trials are largely lacking. Besides, in most trials that are available, efficacy of a drug is judged by reversal of Head up tilt test (HUTT); whether this translates into clinical efficacy is as yet, inconclusive.⁶ Given the various mechanisms underlying the pathogenesis of syncope, it logically follows that ideally, specific therapy should be tailored to specific pathophysiologic mechanisms. However, currently available tools are inadequate to elucidate such mechanisms.

Table III. Common causes of cardiac syncope

Cardiac arrhythmias	Supraventricular tachycardia (SVT), long QT syndrome, Brugada syndrome, Heart Blocks
Outflow tract obstructions	Hypertrophic obstructive cardiomyopathy (HOCM), Aortic Stenosis
Ischemic Heart Disease	Anamolous left coronary artery, coronary aneurysms (post Kawasaki disease)
Myocardial dysfunction	Myocarditis, dilated cardiomyopathy

Cardiac syncope

Cardiac syncope is a serious concern but an uncommon diagnosis. Like in adults, even in pediatrics, sudden death is commonly cardiac in origin, and almost one - fifth of young athletes with sudden death have a history of syncope. Thus, diagnosing cardiac syncope can ultimately be life saving. As stressed earlier, red flags in the history and/or an abnormal physical examination and/or an abnormal ECG suggest cardiac syncope and the child needs appropriate detailed evaluation.¹

Arrhythmias are an important cause of cardiac syncope. Supraventricular tachycardia (SVT) is the commonest symptomatic pediatric dysrhythmia. Though the history may be suggestive (h/o racing heart beat), it is likely to be picked up only when the patient is studied during the episode. Hence, one should be on the lookout for a predisposing abnormality (eg. WPW syndrome). Similarly, one should carefully look for the long QT syndrome; since there are a lot of issues that need to be carefully adhered to in order to enable accurate measurement and calculation, a specialist consultation should be obtained. Brugada syndrome is an inherited disorder of the cardiac sodium channels that increases the susceptibility to fatal ventricular arrhythmias and can be diagnosed by its ECG changes (Table II). While first degree heart block could be an incidental finding in the setting of syncope, second and third degree heart blocks need further evaluation to elucidate the cause. Obstructive cardiac lesions may be suspected if there is a concurrent history of chest pain or exercise induced symptoms, or there are murmurs or other abnormal physical findings. While ischemic heart disease is uncommon in pediatrics, patients with previously undiagnosed Kawasaki disease are at risk (Table III).

Evaluation of suspected cardiac syncope

The further evaluation of a patient with suspected cardiac syncope would depend on the underlying condition that is suspected. If dysrhythmia is suspected, further evaluation may be in the form of ambulatory (Holter) monitoring or loop event monitoring though both have inherent limitations. Occasionally, electrophysiological studies may be required.⁷ Echocardiography can be useful if structural lesions or myocarditis is suspected.

Head Up Tilt Test (HUTT)

Upright tilt testing, if positive, helps to confirm the diagnosis of neurocardiogenic syncope. However, it has its own limitations (reasonable specificity but low sensitivity) and is therefore, not necessary as a routine, if such a diagnosis can be made clinically. Thus, it is recommended

only for those patients who have recurrent unexplained syncope in whom cardiac causes have either been excluded or are not likely. In such cases, a positive HUTT might allay anxiety by labeling a diagnosis.⁷

Routine biochemistry and imaging, if performed in all children with syncope, is rarely useful. Hence, it makes more sense to ask for these tests only if there is some suspicion of a particular disorder or in undiagnosed cases.⁷

Treatment of cardiac syncope

Arrhythmias can be treated with antiarrhythmic drugs, ablation of the arrhythmia origin, or implantable devices like pacemakers or defibrillators, as the case maybe. Brugada syndrome can be treated with placement of an internal defibrillator with an excellent prognosis. When the syncope is due to a structural defect, treatment is aimed at ameliorating the underlying defect.

Summary

Syncope in children is not uncommon. Initial evaluation of any child with syncope should be in the form of a detailed history and physical examination, along with a screening ECG. Though the list of causes is large, vasodepressor syncope is the most common, and is diagnosed by the typical history. It is essentially benign, and its treatment is primarily non pharmacological (education and reassurance). Drug therapy is occasionally required only in cases of recurrent syncope. It is important to exclude potentially life threatening cardiac syncope, which can be suspected by the red flags in the history, and/or an abnormal physical examination and/or abnormal ECG. Additional investigations should be performed only in suspected cases.

Points to Remember

- *A detailed history and a thorough physical examination is the most important initial evaluation in children with syncope*
- *Vasodepressor syncope is the commonest type of pediatric syncope that is diagnosed by its classical history.*
- *Evaluation of pediatric syncope should be able to identify cardiac syncope which is potentially dangerous; an ECG, along with the history and physical examination, has a 96% sensitivity in picking up cardiac syncope.*
- *A long list of aimless investigations may not be useful.*
- *The mainstay of treatment of vasodepressor syncope is largely reassurance and general non*

pharmacologic measures; drugs are only occasionally used in cases of recurrent syncope.

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CLIPPINGS

Clark JD, Dudzinski DM. The culture of dysthanasia: attempting CPR in terminally ill children. Pediatrics. 2013, March.

Both dying children and their families are treated with disrespect when the presumption of consent to cardiopulmonary resuscitation (CPR) applies to all hospitalized children, regardless of prognosis and the likely efficacy of CPR. This “opt-out” approach to CPR fails to appreciate the nuances of the special parent-child relationship and the moral and emotional complexity of enlisting parents in decisions to withhold CPR from their children. The therapeutic goal of CPR is not merely to resume spontaneous circulation, but rather it is to provide circulation to vital organs to allow for treatment of the underlying proximal and distal etiologies of cardiopulmonary arrest. When the treating providers agree that attempting CPR is highly unlikely to achieve the therapeutic goal or will merely prolong dying, we should not burden parents with the decision to forgo CPR. Rather, physicians should carry the primary professional and moral responsibility for the decision and use a model of informed assent from parents, allowing for respectful disagreement. As emphasized in the palliative care literature, we recommend a directive and collaborative goal-oriented approach to conversations about limiting resuscitation, in which physicians provide explicit recommendations that are in alignment with the goals and hopes of the family and emphasize the therapeutic indications for CPR. Through this approach, we hope to help parents understand that “doing everything” for their dying child means providing medical therapies that ameliorate suffering and foster the intimacy of the parent-child relationship in the final days of a child’s life, making the dying process more humane.

ERRATUM

Vol.15, No.1(Jan-March), 2013

Article titled “Changing trends in intravenous maintenance fluid therapy” 2013;15(1):9,

1. Column.2 paragraph.3, last line

- “serious medication errors” – should be read as “serious medication errors can be avoided”

2. Column.2 paragraph.6, line 9

- eg. “If 2000mL/day” – should be read as “If 2400mL/day”

GENERAL ARTICLES

NUTRITIONAL DEFICIENCIES IN NORMALLY GROWING CHILDREN

**Rajeshwari K
**Dubey AP*

Abstract: *Nutritional deficiencies are common in the pediatric population specially among impoverished populations. This is more so among growing children. Overt nutritional deficiencies are well known to all; however subclinical deficiencies in normally growing children are difficult to assess and treat. This article focuses on the common nutritional deficiencies seen in normally growing children with special reference to Indian population.*

Keywords: *Nutritional deficiency, Normal growth, Children*

Nutritional deficiencies have always been a major clinical problem in children. According to the National Family Health Survey (NFHS-3) carried out in 2005-06, child malnutrition rates in India are disproportionately high. The NFHS-3 is the third pan-India survey conducted since 1992 a nationally representative sample of 109041 households. The results are sobering: 46 per cent of children under three are underweight, compared with 28 per cent in Sub-Saharan Africa and 8 per cent in China - another country with an enormous rural poor population. In addition to the 46 per cent who are underweight, 39 per cent are stunted, 20 per cent wasted and 80 per cent anemic. More than 6,000 Indian children below the age of five die every day due to malnourishment or lack of basic micronutrients such as iron, iodine, Vitamin A, zinc or folic acid.

The classic forms of many of the well known nutritional deficiencies are frequently described in medical literature, but many nutritional deficiencies can occur in otherwise normally growing children who are asymptomatic. The two most common deficiencies seen in children who are growing normally are iron and vitamin D deficiencies.

These deficiencies are surprisingly common and can cause significant morbidity. This article describes these nutritional deficiencies and other less commonly seen deficiencies in children who are otherwise growing normally.

Iron deficiency

Iron deficiency and iron-deficiency anemia affects 2.4 million and 490 000 US children, respectively.^{1,2} Children 1 to 3 years old are particularly vulnerable, because maternal iron stores are depleted during a period of rapid growth.^{2,3} A recent analysis of the National Health and Nutrition Examination Survey (NHANES) III found the prevalence rates of iron deficiency and iron-deficiency anemia among US toddlers to be 9% and 3%, respectively².

Iron-deficiency anemia in infancy and early childhood is associated with behavioral and cognitive delays, including impaired learning, decreased school achievement, and lower scores on tests of mental and motor development.⁴ Given the detrimental long-term effects and high prevalence of iron deficiency, its prevention in early childhood is an important public health issue.

Effective approaches to the prevention of iron deficiency in infancy and early childhood should include screening and counseling practices targeting children identified to be at high risk for iron deficiency. Iron deficiency affects 20% to 25% of infants worldwide, and most studies of risk factors for iron deficiency in early childhood have been conducted in Latin America, Africa, India, Europe, and Canada. Several studies have demonstrated a high prevalence of iron deficiency in the United States among low-income infants and children, who may experience food insecurity and have diets low in iron. Important dietary risk factors include exclusive breastfeeding beyond 6 months not supplemented by iron-rich foods or vitamins with iron, early introduction of milk, prolonged bottle feeding, and excessive cow's milk consumption. An association between maternal prenatal anemia and iron deficiency has also been reported.

In India among children aged 6 to 35 months, there is a noticeable variation in the prevalence of moderate IDA (1998/99) by demographic and socioeconomic characteristics. It tends to be higher among children from

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disadvantaged groups, namely those living in rural areas, poor households, and from scheduled castes/tribes. The prevalence of mild and severe anemia is less variable, however, hovering around 23% and 5%, respectively. There is almost no difference in the prevalence of IDA by children's gender.

Iron deficiency in industrialized countries presents most commonly as a mild-to-moderate microcytic, hypochromic anemia in an asymptomatic, well-nourished infant. Uncommon but also reported are infants with severe anemia who present with pallor, lethargy, irritability, poor feeding, and cardiomegaly. Although typically presenting as a nutritional anemia, IDA may present secondary to other diseases including celiac disease, *Helicobacter pylori* infections, and the anemia of chronic disease. Pica (the craving for substances largely non-nutritive such as clay or paper products) and pagophagia (craving for ice) are common features associated with ID. It may be present in children who are not anemic and will respond rapidly to treatment with iron, often before any increase is noted in the hemoglobin concentration.

For infants/children presenting with a mild microcytic, hypochromic anemia with a presumptive diagnosis of ID anemia, the most cost-effective strategy is a therapeutic trial of iron. Several laboratory tests can be helpful in confirming the diagnosis of ID anemia. Although an elevated red cell distribution width is the earliest hematologic manifestation of ID, ID in infants and young children is usually identified by a serum ferritin concentration of less than 12 ng/mL. IDA is diagnosed by a low hemoglobin concentration in conjunction with a low serum ferritin. A more complete evaluation for IDA would also include a serum iron, total iron-binding capacity, and transferrin saturation. Other laboratory tests, although not routinely used, such as erythrocyte protoporphyrin, serum transferrin receptor, and reticulocyte hemoglobin content, may prove useful tools for measuring ID.⁵

For infants with confirmed IDA, ferrous sulfate (3-4 mg/kg of elemental iron, in divided doses, between meals with a citrus juice) is the standard of care. Ferrous sulfate at 3 mg/kg should produce an increase of greater than 1 g/dL per week in patients with ID. Iron absorption is increased if the ferrous sulfate is given with juice rather than milk. Iron should be continued in responders for 2 to 3 months after normalization of hemoglobin values to replace iron stores. If the patient fails to respond after 4 weeks of therapy, a review of the patient's history should take place for medication dosing and administration errors, appropriate dietary modifications, or history of a recent illness. Other laboratory studies, including a serum ferritin

level, should be obtained to evaluate the anemia further and to rule out conditions simulating (ie, thalassemias and the anemia of chronic disease) or complicating (eg. concomitant vitamin B12 or folic acid deficiency) ID anemia. Close follow-up should occur after appropriate treatment to ensure the patient's response to iron therapy.

Stroke and iron deficiency anemia

Previously healthy children with stroke were 10 times more likely to have iron-deficiency anemia than healthy children without stroke.⁶ Furthermore, children with iron-deficiency anemia accounted for more than half of all stroke cases in children without an underlying medical illness, which suggests that iron-deficiency anemia is a significant risk factor for stroke in otherwise healthy young children. Primary prevention and early identification of iron-deficiency anemia must remain a priority. To conclude healthy breastfed baby is protected against manifest iron deficiency anaemia during the first few months of life, although some susceptible infants may develop iron deficiency. The finding of iron deficiency anaemia in 6% of formerly breastfed infants at 7 or 10 months of age who had been fully breastfed for the first 4 months of life supports the recommendation of a timely introduction of iron-rich complementary foods in breastfed infants.⁷

Vitamin D deficiency

Vitamin D is a prohormone that is essential for the normal absorption of calcium in the gastrointestinal tract. Deficiency in vitamin D leads to hypocalcemia and hypophosphatemia with resultant rickets in children and osteomalacia in adults. In adults, vitamin D deficiency has been linked to cardiovascular disease, insulin resistance, and hypertension.

The resurgence of vitamin D deficiency is likely a result of several dietary and environmental factors, including body mass index, milk ingestion, and sun exposure. In nonindustrialized nations vitamin D deficiency remains a major public health problem. Vitamin D deficiency causes nutritional rickets. The primary abnormality may be dietary deficiency or decreased vitamin D activity, which leads to a decrease in intestinal absorption of calcium. Although the majority of pediatric patients with low vitamin D level are asymptomatic, some may develop secondary hyperparathyroidism and characteristic changes in the growth plates and metaphyseal bones.

The vitamin D status of an infant depends upon the amount of vitamin D transferred from the mother prenatally and on the amount of vitamin D ingested or produced by the skin during exposure to ultraviolet light postnatally.

Maternal-fetal transfer of vitamin D is mostly in the form of calcidiol (25-OH vitamin D), which readily crosses the placenta. The half-life of calcidiol is approximately 3 to 4 weeks. Thus, the serum concentration of vitamin D falls rapidly after birth unless additional sources are available.

In North America, infant formula, cow's milk, and cereals are fortified with vitamin D. All infant formulas in the United States contain at least 400 IU/L of vitamin D. Nonetheless, the diet of most breast-fed infants and many formula-fed infants does not provide the recommended intake of vitamin D. Vitamin D deficiency rickets commonly presents between 3 months and 3 years of age, when growth rates (and calcium needs) are high, and exposure to sunlight may be limited. The main reasons for inadequate vitamin D supply in infants from Western countries are prolonged breast-feeding without vitamin D supplementation, and concomitant avoidance of sun exposure. The recommended intake of vitamin D to prevent deficiency in normal infants and young children is 200 to 400 IU/day. Human milk typically contains less than 25 IU of vitamin D per liter. Dark skin is an additional risk factor for developing rickets in breast-fed infants as dark-skinned individuals produce less vitamin D in response to sunlight. The vitamin D concentration of the breast milk of dark-skinned mothers is less than that of lighter-skinned individuals. In high-risk populations, most mothers of breast-fed infants with rickets are deficient in vitamin D. Despite a clearer understanding of predisposing factors and attempts at preventative strategies, nutritional rickets has made a surprising resurgence in many parts of the world. Recent reports have not only come from temperate regions with limited sunshine such as Canada, New Zealand, the UK, and USA, but also from sunnier climates such as Australia, Ethiopia, and Saudi Arabia. A dramatic increase in the presentation of vitamin D deficient rickets to major paediatric centres in Sydney, a large, modern city with good nutritional and health standards and relatively high sunlight hours has recently been reported.⁸

A recent study from New Delhi concluded that hypovitaminosis D in lactating mothers is strongly correlated with hypovitaminosis D in neonates and infants. Given this correlation, infants born to vitamin D deficient mothers are prone to develop hypocalcemic seizures.⁹ Another study from North India showed the unexpectedly high prevalence of hypovitaminosis D among pregnant women. The physiologic relevance of the finding is substantiated by the negative correlation with PTH and the positive correlation with cord blood 25(OH) D.¹⁰

Another study from New Delhi showed high

prevalence of vitamin D deficiency and insufficiency in breastfed infants and their mothers, with radiological rickets in a third of infants with 25OHD < 10 ng/ml.¹¹

The most widely used treatment for vitamin D deficiency is vitamin D₂ (ergocalciferol). Infants younger than 1 month with vitamin D deficient rickets should receive 1000 IU daily, infants aged 1 to 12 months should receive between 1000 and 5000 IU daily, and children older than 1 year should receive 5000 IU daily. Treatment is continued at these doses until radiographic evidence of healing is observed. The dose of vitamin D is then reduced to 400 IU daily. Radiographic evidence of healing usually occurs after 3 months of treatment. Calcium intake should be maintained at approximately 1000 mg per day to avoid the so-called hungry bone syndrome (worsening hypocalcemia after the start of vitamin D therapy). This is usually accomplished by administering supplements of 30 to 75 mg/kg of elemental calcium per day in three divided doses.

An alternative treatment protocol is the so-called Stoss therapy, which consists of a oral vitamin D (600,000 IU) given on a single day.¹² This amount of vitamin D approximately corresponds to a 3-month course of 5000 IU per day and should be sufficient to induce healing within 3 months. Stoss therapy may be advantageous when compliance with therapy or follow-up is a problem. However, such high doses of vitamin D can lead to hypercalcemia. Doses of 150,000 or 300,000 IU appear to be equally effective, but with less risk of hypercalcemia.

Calcium Deficiency

Nutritional rickets remains prevalent in many parts of the world. Because ample sunlight exists in many of the countries where the incidence of rickets is high, researchers have suggested that insufficient calcium intake rather than primary vitamin D deficiency may be the main causative factor. Most of the children in these studies had normal serum 25-OH vitamin D concentrations and high serum 1, 25-OH₂ vitamin D concentrations, indicating adequate intake of vitamin D. A randomized, double-blind, controlled trial of 123 Nigerian children with rickets showed that baseline intake of calcium was very low (about 200 mg/d).¹³ These children responded better to treatment with calcium alone or in combination with vitamin D than to treatment with vitamin D alone.

Although vitamin D and iron are the most common nutritional deficiencies in normally growing infants and children, an array of other nutritional deficiencies can occur in specific clinical situations. These occur in certain diseases and in special diets, such as vegetarian/vegan diets.

Table I. Prevalence of micronutrient deficiencies in south asia

	Iron deficiency				Vitamin A deficiency			Iodine deficiency			Folate deficiency
	IDA in children <5y (%)	IDA in women 15-49y (%)	IDA in pregnant women (%)	Maternal death from severe anemia/yr (no.)	Child deaths precipitated (no.)	Children <6 w/ subclinical VAD (%)	Children <6 w/ clinical VAD (%)	Children born mentally impaired (no.)	Total Goiter Rate (TGR) (%)	Total Goiter Rate (TGR) in school children (%)	Neural tube defects (no.)
Afghanistan	65	61	-	-	50,000	53	-	535,000	48	-	2,250
Bangladesh	55	36	74	2,800	28,000	28	0.7	750,000	18	50	8,400
Bhutan	81	55	68	<100	600	32	0.7	-	-	14	150
India	75	51	87	22,000	330,000	57	0.7	6,600,000	26	19	50,000
Nepal	65	62	63	760	6,900	33	1.0	200,000	24	40	1,600
Pakistan	56	59	-	-	56,000	35	-	2,100,000	38	-	11,000
South Asia Region Total				25,560	471,500			10,185,000			73,400
World Total				50,000	1,150,000			19,000,000			204,000

Source: UNICEF 2003b; WHO 2000; UNICEF and MI 2004a

Less common causes of nutritional deficiencies in normal growing infants and children are reviewed in the following paragraphs.

Vitamin B Complex Deficiency

Beriberi in infants becomes clinically apparent between the ages of 2 and 3 months. The clinical features are variable and may include a fulminant cardiac syndrome with cardiomegaly, tachycardia, a loud piercing cry, cyanosis, dyspnea, and vomiting.

Beriberi is an endemic disease among the Naporuna indigenous people of Ecuadorean Amazonia (annual morbidity rate of 1.5%). Although the diagnosis of the disease was made clinically, an effective and quick response to thiamine treatment excluded other differential diagnoses, such as tropical ataxic neuropathy. Although indigenous people have several possible dietetic risk factors, none of them adequately explain the disease's high incidence.¹⁴

In the United Kingdom, the National Diet and Nutrition Survey reported a high prevalence of poor riboflavin status in adolescent girls and young women. It has been reported

that 95% of adolescent girls (15–18 years) and 75% of young women (19–24 years) in the United Kingdom have poor riboflavin status as measured by the erythrocyte glutathione reductase activation coefficient assay. Most cases of riboflavin deficiency go undetected because of the mild nature and nonspecific signs and symptoms of the deficiency. Significant deficiency is characterized by sore throat, hyperemia of pharyngeal mucous membranes, edema of mucous membranes, cheilitis, stomatitis, glossitis, normocytic-normochromic anemia, and seborrheic dermatitis.

Niacin deficiency is occasionally reported in children on very poor diets. The exact subclinical deficiency in India is still unknown. Pyridoxine deficiency is seen classically in one of six pyridoxine-dependent syndromes: pyridoxine-dependent seizures, B6 responsive anemia, xanthurenic acidemia, cystathionemia, homocystinuria, and type 2 hyperprolinemia. Unusual as an isolated nutritional deficiency, pyridoxine deficiency has been associated with isoniazid treatment and in exclusively breast-fed infants older than 6 months.

Various micronutrient deficiencies seen in South Asian Countries is summarized above in Table 1.

The prevalence of micronutrient deficiencies among children and women of reproductive age in India is also consistently among the highest in the world.¹⁵ For example, the prevalence of iron deficiency anemia (IDA) among preschool children is over 75%; although the nationwide prevalence of clinical Vitamin A deficiency (VAD) is less than 1-2%, up to 60% of preschool children have subclinical VAD; and, about one in four school children have goiter, a sign of severe iodine deficiency.

The prevalence of Vitamin A deficiency (VAD) in India is one of the highest in the world, especially among preschool children. The prevalence of subclinical VAD ranges from 31% to 57% among preschool children and a further 1% to 2% of children suffer from clinical VAD. With its large population, India is home to more than a quarter of the world's population of preschool children suffering from subclinical VAD (35.4 out of 127.3 million) and a third of the preschool population with xerophthalmia (1.8 million out of 4.4 million). As a result of this high prevalence, VAD is estimated to precipitate the deaths of more than 0.3 million children annually in India. . Clinical VAD is a severe form of Vitamin A deficiency, resulting in xerophthalmia, symptoms of which include night blindness, Bitot's spots, xerosis, and keratomalacia. If not treated early enough, it can eventually leads to blindness. Subclinical VAD, defined by a serum retinol concentration of less than 0.7 $\mu\text{mol/L}$, is associated with increased vulnerability to a variety of infectious diseases and, therefore, an increased risk of mortality and morbidity.

Iodine deficiency disorders (IDD)

Although the prevalence of iodine deficiency disorders (IDD) in India is lower than in most South Asian countries, the problem is ubiquitous and affects millions of people. One survey showed that more than 85% of districts (241 out of 282) are endemic for IDD. This places about 329 million people at risk, equivalent to a third of India's population or a sixth of the total global population that is at risk of IDD. Of those who suffer from IDD in India, 51 million are school-aged children (aged 6 to 12 years). A third of all children in the world that are born with IDD-related mental damage live in India.

Points to remember

- *Nutritional deficiencies are common in children.*
- *In normally growing children it is important to diagnose various deficiencies specially when manifestations of deficiencies are not overt.*

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GENERAL ARTICLES

ANTIEMETICS IN PEDIATRICS

***Jeesson C Unni**

Abstract: *A consensus on the indications for the use of antiemetics in children needs to be formulated as most drugs under this drug class have side effects and most common causes of vomiting in children do not require medication for control of emesis. This article is an attempt to resolve certain issues related to the use of antiemetics in children.*

Keywords: *Antiemetics, Cytotoxic drugs, Chemotherapy, Post operative vomiting, Acute gastroenteritis, Motion sickness, Phenothiazines, 5-HT₃ antagonists.*

Antiemetics are medications that are used to prevent or relieve nausea and vomiting. Vomiting occurs due to postoperative reaction to anesthesia, gastroenteritis, influenza, radiotherapy, chemotherapy, uremia and hepatitis. This can result in dehydration and severe negative health outcomes for patients experiencing these symptoms. Antiemetics are used in these situations in addition to the specific management of the underlying illnesses. More often than not, doctors who prescribe antiemetics for children are doing so off-label¹.

Principles of use of antiemetics²

1. Prescription of antiemetics should not delay the diagnosis. It is important to simultaneously evaluate for the cause.
2. In certain conditions antiemetics are unnecessary and sometimes harmful. Eg. Diabetic ketoacidosis, digoxin and antiepileptic overdose.
3. In conditions where drug treatment is indicated for control of nausea and vomiting, drug appropriate for the etiology needs to be chosen.

Pharmacological groups: Though antiemetics are generally classified as antihistamines, (eg. buclizine,

cyclizine, dimenhydrinate, diphenhydramine, hydroxyzine hydrochloride) Phenothiazines (eg. chlorpromazine, prochlorperazine, promethazine and 5-HT₃ receptor antagonist (eg. Ondansetron, dolasetron and granisetron) a practical classification is given below.

1. Less frequently used drugs: Antihistamines, phenothiazines and metoclopramide. They are no more preferred as antiemetics of first choice because of various side effects like CNS depression, dystonia and others and also due to availability of safer and effective drugs. But antihistamines still find a place in the management of motion sickness.

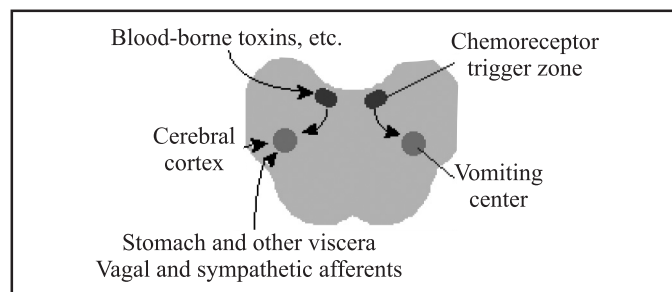
2. Most commonly used drug at present: 5-HT₃ receptor antagonists eg. Currently ondansetron is the most preferred antiemetic drug. Dolasetron and granisetron are the newer generation drugs in that class.

3. Drug used as second line : eg. Dexamethazone. This is used along with ondansetron for delayed vomiting induced by chemotherapy and radiotherapy by oncologists

4. Third line drugs: **Nabilone** a synthetic cannabinoid and aprepitant are used as third line drugs, mostly in adults. Sometimes they are used as third line drugs in children, mainly in chemotherapy induced vomiting.

Drug classes, mechanism of action and recommended indications:

All the drugs act through the vomiting centre in the medulla. This centre receives input from four different sources. 1. Gastrointestinal tract (GIT) 2. Vestibular apparatus 3. Chemoreceptor Trigger Zone (CTZ) and 4. Cerebral cortex. Proper understanding of the pathophysiology of vomiting, helps us to tailor the therapy appropriate for the problem.



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These four sources through two pathways - one neural and one humoral - lead to activation of vomiting center which makes the ultimate decision, based on input from various sources, among whom the chemoreceptor trigger zone is more influential.

Phenothiazines: These are considered for the prophylaxis and treatment of nausea and vomiting associated with general anesthetics³ neoplastic disease and radiation sickness. However 5-HT₃ antagonists are preferred in these conditions⁴ because of severe dystonic reactions of phenothiazines.⁵ Both haloperidol and droperidol, a butyrophenone, structurally related to it, block dopamine receptors in the chemoreceptor trigger zone. This central antiemetic action prevents postoperative nausea and vomiting to some extent^{6,7} Haloperidol is also used orally or by continuous intravenous or subcutaneous infusion for most metabolic causes of vomiting (eg. hypercalcemia, renal failure)⁸ and for relief of nausea in palliative care.

Metoclopramide: This is an antiemetic and prokinetic and is superior to phenothiazines. But 5-HT₃ receptor antagonists may be more efficacious and less toxic.⁴

Domperidone: This has the advantage over metoclopramide and phenothiazines as it causes less central effects such as sedation and dystonic reactions because it does not readily cross the blood-brain barrier. Domperidone is also used to treat vomiting due to contraceptives. It may be also used for preventing and treating emesis associated with chemotherapy.

5HT₃-receptor antagonists: Ondansetron and granisetron block 5-HT₃ receptors in the gastro-intestinal tract and in the CNS. They are of value in the management of nausea and vomiting in children receiving cytotoxics^{4,9} and in postoperative nausea and vomiting.^{9,10} Antiemetics are generally avoided in acute gastroenteritis. However if vomiting is very severe ondansetron can be used along with ORT¹¹. Its use could decrease the need for hospitalisation and administration of IV fluids in a child with gastroenteritis.

Nabilone This drug is a synthetic cannabinoid with antiemetic properties. Because of medical and legal concerns they are not 1st line drugs but may be tried for nausea and vomiting caused by cytotoxic chemotherapy that is unresponsive to conventional antiemetics and in

Table I. Input sources, neurotransmitters and drugs acting on various sites

Input sources	Stimulus	Neurotransmitters	Drugs acting in this site
1.Gastrointestinal tract	Distension or inflammation	Serotonin, acetylcholine, histamine and substance P	5-HT ₃ receptor antagonists block serotonin stimulation in the periphery and in CTZ .
2.Vestibular apparatus	Motion sickness	Stimulus of the vestibular apparatus is mediated through histamine and acetylcholine receptors.	Antihistamines – Used to treat motion sickness.
3.CTZ	The CTZ senses rapidly rising chemicals in the blood	Dopamine, acting on D ₂ receptors, and serotonin, acting on 5HT ₃ receptors.	Phenothiazines, domperidone metoclopramide and droperidol block dopaminergic receptors in the CTZ
4.Cerebral cortex	Pathways are less well understood Anticipatory nausea and vomiting through olfactory and emotional stimuli	The cerebral cortex and limbic system modulate complex experiences such as taste, sight and smell as well as memory (involved in anticipatory nausea) and emotion.	

children with breakthrough vomiting¹². Side-effects such as drowsiness and dizziness occur frequently with standard doses.

Dexamethasone: It is used as a second line drug when ondansetron is ineffective or in delayed vomiting that occur beyond 24 hrs and sometimes up to 7 days following chemotherapy.. Dexamethasone, given orally, is the drug of choice for preventing delayed cytotoxic-induced nausea and vomiting; it is used alone or with metoclopramide⁴.

Antihistamines: They are mainly used in the management of motion sickness. Nausea in the first trimester of pregnancy is generally mild and does not require drug therapy. On rare occasions if vomiting is severe, short-term treatment with an antihistamine, such as **promethazine**, may be required.¹³

Choice of antiemetics in various clinical situations

Vomiting in common illnesses: Ondansetron is the ideal choice in the relief of nausea and vomiting in common conditions such as viral fever and gastroenteritis.

Post operative nausea and vomiting: Severity depends on type of anesthetics and type and duration of surgery. Female gender, past history of vomiting in the post operative period or motion sickness and intraoperative or postoperative use of opioids increase the risk of vomiting. Ondansetron should be the first choice and if this fails two or more classes of antiemetics have to be used. Eg. Dexamethazone

Chemotherapy /radiotherapy induced vomiting: It is classified into three types acute (vomiting developing within 24 hrs of therapy), delayed (if it occurs after 24 hours) and anticipated (if child develops vomiting prior to subsequent therapy). Delayed and anticipatory vomiting is difficult to control than acute and they require more than one drug of different mechanism of action. eg. Ondansetron.or metoclopramide with steroids and phenothiazines.

Motion sickness: Antiemetics should be given prophylactically to children with motion sickness rather than after nausea or vomiting develop. The most effective drug for the prevention of motion sickness is the transdermal hyoscine patch that needs to be applied several hours before travel¹⁴. The sedating antihistamines like promethazine may be given to prevent motion sickness. However less sedating antihistamines such as cyclizine or cinnarizine are preferred. Both are less effective than hyoscine but are better tolerated.

Individual drugs¹⁵

Prochlorperazine

Dosage: Oral/rectal > 1yr 0.1-0.25 mg/kg/dose 2-3 times daily. Deep IM 0.1-0.15 mg/kg/24hr divided in 2-3 doses.

Contra-indicated in pregnancy. Not recommended for use in children less than 10kg or one year of age. Reduce dose in renal impairment. Caution in liver disease (may precipitate coma). Avoid in patients with epilepsy. Prochlorperazine has been associated with dystonic reactions particularly after a cumulative dosage of 500 micrograms/kg; it should be used cautiously in children.

Side-Effects: Nasal stuffiness, dry mouth, insomnia and agitation. Extrapyramidal symptoms (especially in seriously ill children, those less than 10kg or one year of age). May rarely cause neuroleptic malignant syndrome. Respiratory depression may occur in susceptible patients. Postural hypotension is yet another side effect.

Drug-Interactions: Increased extrapyramidal side-effects with metoclopramide and lithium. Decreased effect of antiepileptics. Decreased levels with antacids. Enhanced sedative effects with anxiolytics and hypnotics. Avoid with desferrioxamine.

Poisoning: Symptoms include drowsiness or loss of consciousness, hypotension, tachycardia, ECG changes, ventricular arrhythmias and hypothermia. Severe extrapyramidal dyskinesias may occur.

Chlorpromazine

Dosage: Nausea and vomiting of terminal illness - Oral - 2-12yr 500microgm/ kg/dose (max 40mg/day; 12-18yr 10-25mg/dose (max 75mg/ day) - 4 times daily.

Contra-Indications: Bone marrow depression; pheochromocytoma. Pregnancy: Inadequate evidence of safety in human pregnancy but it has been widely used for many years without apparent ill consequence. It may occasionally prolong labour and at such a time should be withheld until the cervix is dilated 3-4cm. possible adverse effects on the neonate include lethargy or paradoxical hyper excitability, tremor and low Apgar score. Avoid unless considered essential. Breast Feeding: Excreted in breast milk: can cause drowsiness in baby. Breast-feeding should be suspended during treatment.

Side-Effects: Drowsiness and sedation, antimuscarinic effects, hypotension, tachycardia, cardiac dysrhythmias, extrapyramidal symptoms and neuroleptic malignant syndrome, tardive dyskinesia after prolonged administration, hypothermia, gynecomastia, galactorrhea, impotence, menstrual disturbances, weight gain, lowered threshold for seizures, sensitivity reactions; bone marrow depression, leucocytosis, hemolytic anemia, contact hypersensitivity, photosensitivity, rashes, cholestatic jaundice, corneal and lens opacities and skin pigmentation with prolonged use.

Drug-Interactions: Plasma concentration increased by propranolol; enhances antimuscarinic effects of drugs; extrapyramidal effects increased by lithium.

Poisoning: General sedation and depression of consciousness and respiration, hypothermia and tachycardia; consequent hypoxia and acidosis; seizures; dystonic reactions respond to antimuscarinic drugs, particularly benzatropine (benztropine) and procyclidine

Trifluoperazine

Dosage: Vomiting not responding to other medications - Oral - 3-5yr up to 1mg/day in div doses; 6-12yr up to 4mg/day in div doses; 12-18yr 2-4mg/day in div doses (max 6mg).

Haloperidol

Dosage: Nausea - 12-18 yr 0.5-2mg 2-3 times daily.

Metoclopramide

Dosage: Oral, IM or slow IV < 12yr 0.3 mg/kg/day (max 10gm/day) in 3 divided doses. 12-18 yr 5-10mg /dose 3 times daily. IV: as a slow bolus injection over at least 5 minutes or for doses over 10mg at a suggested concentration of 200 micrograms per ml over 15-30 minutes. Dilute in NaCl 0.9% or glucose 5% if required. For diagnostic procedures, IV dose is given 10-20 minutes before procedure. In mild to moderate renal impairment use 75% of dose and in severe impairment 25-50% of dose. Reduce dose in severe liver disease.

Side-Effects: Extrapyramidal side-effects, dystonic reactions including oculogyric crises; more common in children and young adults. Treat with an anticholinergic eg. benzatropine (benztropine), procyclidine. Drowsiness, diarrhea, restlessness.

Drug-Interactions: Antimuscarinics and opioid analgesics antagonize the effect of metoclopramide on gastrointestinal activity. Paracetamol and aspirin give an enhanced effect. Antipsychotics increase risk of extrapyramidal effects.

Domperidone

Dosage: Gastroesophageal reflux and gastric stasis - Oral 1 month-12yr 200-400 microgm/kg/dose 3-4 times daily before food and at night. Not often used < 2 yr. Radiotherapy / chemotherapy induced vomiting - Oral 1 month -12 yr 200-400 microgm/kg as single dose. Could give once in 4 - 8hr.

Contra-Indications: Less effective in centrally-mediated nausea and vomiting than metoclopramide. Prolonged QT

intervals have been seen in neonates. It is recommended in neonates to check ECG prior to dosing and at regular intervals afterwards (2weeks after starting). Pregnancy: avoid if possible.

Side-Effects: Acute dystonic reactions (less common than with metoclopramide). Occasional rashes.

Drug-Interactions: May enhance the absorption of concurrently administered drugs. Actions may be antagonized by antimuscarinics and opioid analgesics.

Ondansetron

Dosage: Nausea and vomiting - Chemotherapy induced - IV < 12 yr 5mg/m² and 12-18 yr 8mg as single dose just before chemotherapy and repeated 8-12 hrly during therapy and for 24 hr after. Oral following initial IV dosing to get control - < 12 yr 4mg and 12-18 yr 8mg 2-3 times daily for up to 5 days after a course of chemotherapy. Post operative nausea and vomiting - IV slow infusion 2-12 yr 0.1mg/kg (max 4 mg) and 12-18yr 4mg. Pruritis - Oral 2-12 yr 2-4mg/dose and 12-18 yr 4-8 mg/dose 2 times daily. Palliative for nausea/vomiting and pruritis: IV/SC 5mg/m² continuous over 24 hr.

Administration: Oral: the melt tablets melt in the mouth; no water required, but they must be swallowed (not buccally absorbed). IV: injection over 2-5 minutes; infusion over 15 minutes diluted in glucose 5% or NaCl 0.9%. NB. In palliative care ondansetron has been administered as a 24 hourly Sc or IV infusion.

Contra-Indications: Hypersensitivity to any component of the preparation.

Pregnancy: use only if expected benefit to patient outweighs potential risk to fetus.

Breast Feeding: Not recommended; ondansetron passes into breast milk.

Side-Effects: Headache, constipation.

Drug-Interactions: No specific interactions reported.

Granisetron

Dosage: Oral - 1mth -12yr - 20microgm/kg/dose to a max of 1mg; 12-18yr - 1mg administered 1-2 times with the first dose given 1hr prior to starting chemotherapy. IV - 1mth - 12yr - 40microgm/kg/dose to a max of 3mg; 12- 18yr - 3mg as a single dose just before administering chemotherapy. One more dose may be given within a 24hr period.

Administration: Injection or infusion given over 5 min in 10-30mL normal saline or 5% dextrose.

Contra-Indications: Hypersensitivity to the drug. In hepatic impairment the dose may need to be reduced.

Pregnancy: No teratogenic effect in animals. Avoid if possible due to lack of data in pregnancy.

Side-Effects: Headache, constipation.

Drug-Interactions: None described.

Dexamethasone

Dosage: Anti-emetic with chemotherapy - IV/oral per dose < 1yr 250 microgram-1 mg, 1-5 yr 1-2 mg, 6-12 yr 2-4 mg and 12-18 yr 4 mg thrice daily until 48 hr after chemotherapy.

Administration: Tablets will disperse in water. IV: can be given as a bolus over 3-5 minutes. The injection can be diluted in NaCl 0.9% or glucose 5%.

Promethazine

Dosage: Vomiting and nausea - Oral 1mg/kg/24hr in 4 divided doses.

Cyclizine

Dosage: Children - 6-12yr orally 25mg/dose up to 3 times/24hr as required and older Adolescents 50mg up to 4th - 6th hrly 30 min before travel (max 200mg/24hr; IM 50mg 4-6 times. Administration: IV: by slow IV injection over 3-5 minutes. May be diluted if required to a maximum 1:1 dilution with water for injection only. Can be given as a continuous IV infusion. Orally: tablets may be given crushed and dispersed in water. SC: occasionally associated with irritation at site of continuous SC infusion. If affected consider increasing the dilution of drug in the infusion or adding hyaluronidase or low dose hydrocortisone to the syringe. Do not mix with any solution containing chloride ions as this will lead to precipitate formation.

Contra-Indications: Sedation. Cyclizine should be used with caution in patients with glaucoma, obstructive disease of the gastrointestinal tract and hepatic disease (may induce coma). Dose reduction may be necessary in renal impairment. Caution may be required in epilepsy and severe heart failure. Cyclizine injection may have a hypotensive effect.

Side-Effects: Drowsiness, occasional dry mouth and blurred vision. Headache, psychomotor impairment and antimuscarinic effects such as urinary retention and

gastrointestinal disturbance. Urticaria, drug rash, tachycardia, restlessness, nervousness, insomnia and hallucinations.

Drug-Interactions: There may be additive effects with the co-administration of CNS depressants. May enhance the side-effects of other anticholinergic drugs. Increased antimuscarinic and sedative effects with tricyclics, antimuscarinics, anxiolytics and hypnotics.

Poisoning: Symptoms of acute toxicity arise from peripheral anticholinergic effects and effects on the central nervous system including ataxia, hyperkinesias, extrapyramidal motor disturbance, convulsions and respiratory depression. The incidence of convulsions in children <5 years of age is about 60% when oral dose ingested exceeds 40mg/kg.

Cinnarizine

Dosage: Vestibular disorders - Oral - 5-12 yr - 15mg 3 times daily, 12-18yr 30mg 3 times daily. Motion sickness - Oral - 5-12 yr - 15mg 2hr before travel then 7.5mg 8th hrly during journey if necessary, 12-18yr 30mg 2hr before travel then 15mg 8th hrly during journey if necessary.

Contra-Indications: Hypersensitivity to the drug. Avoid during work requiring concentration.

Pregnancy: Use with caution. Breast feeding: Use with caution

Side-Effects: Dry mouth, sedation, decreased alertness and concentration, fatigue, epigastric discomfort, urinary hesitancy and headache.

Drug-Interactions: Barbiturates, analgesics will enhance CNS depression.

Points to Remember

- *Prescribe an antiemetic only when the cause of vomiting is known, as it might otherwise delay diagnosis.*
- *In certain conditions antiemetics are unnecessary and sometimes harmful when the cause can be otherwise treated such as in diabetic ketoacidosis, or in digoxin or antiepileptic overdose.*
- *In conditions where drug treatment is indicated for control of nausea and vomiting, drug appropriate for the etiologic needs to be chosen.*

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CLIPPINGS

Meropol SB, Localio AR, Metlay JP. Risks and benefits associated with antibiotic use for acute respiratory infections: a cohort study. Ann Fam Med. 2013 March

Antibiotics are frequently prescribed for acute nonspecific respiratory infections (ARIs), presumably to avoid small risks of progression to serious bacterial illness. However, even low risks of associated adverse drug events could result in many such events at the population level. Our objective was to assess the risks and benefits of antibiotic use in a cohort of patients with ARIs, comparing outcomes of patients who were prescribed antibiotics with outcomes of patients not receiving antibiotics. Compared with patients with ARI who were not treated with antibiotics, patients who were treated with antibiotics were not at increased risk of severe adverse drug events and had a small decreased risk of pneumonia hospitalization. This small benefit from antibiotics for a common ambulatory diagnosis creates persistent tension; at the societal level, physicians are compelled to reduce antibiotic prescribing, thus minimizing future resistance, whereas at the encounter level, they are compelled to optimize the benefit-risk balance for that patient.

DERMATOLOGY

IMMUNOBULLOUS DISEASES IN CHILDREN

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Abstract: *Auto immune blistering disorders are heterogeneous group of diseases that result from auto antibodies generated against target antigens found in the skin and mucous membranes. This process leads to a variety of disruptions in keratinocyte adhesion and cellular integrity, resulting in fluid accumulation and development of blisters. Physicians should have an appreciation and understanding of autoimmune blistering disorders in the pediatric population when formulating a differential diagnosis of a patient who presents with skin blistering. Early detection and discrimination between the varied autoimmune blistering disorders can change the course of treatment and outcome. Due to the similarity in clinical presentation among different diseases within this category, histopathologic evaluation and, especially, immunofluorescence studies are necessary to establish the definitive diagnosis.*

Keywords: *Immunobullous diseases, Children*

Immuno bullous diseases are autoimmune disorders that affect the skin and mucous membranes of the mouth, eyes, or genital areas. Immuno bullous disorders occur when the body's immune system errantly makes antibodies to the skin proteins. In the skin, there are proteins that attach epidermal cells to each other and proteins that attach the epidermis to the dermis. These proteins are the glue that keep the skin intact. In other words they maintain coherence between epidermal cells and attachment between the dermis and epidermis. When these proteins are damaged by auto immune process, the cells separate from each other or from the underlying dermis and a vesicle develops.

For convenience the Immuno bullous diseases can be broadly divided into two main groups, according to the level of cleavage, the epidermal and the dermal.

The following are the common Immuno bullous disorders seen in children

The epidermal diseases include:

- Pemphigus vulgaris
- Pemphigus foliaceus
- Paraneoplastic pemphigus.

The dermal diseases

- Linear Ig A dermatosis (Chronic bullous dermatosis of childhood)
- Childhood bullous pemphigoid
- Bullous lupus erythematosus

The rarer immune bullous disorders are

- Neonatal pemphigus
- IgA pemphigus
- Mixed Immuno bullous disease
- Dermatitis herpetiformis
- Epidermolysis bullosa aquisita

Epidermal immunobullous disorders¹

Pemphigus vulgaris (PV)

This is the commonest form of epidermal immunobullous disorder. Children under 10 years of age are extremely rare implying an increase in incidence during puberty. The youngest female reported was 4 years old, while the youngest male was 3 years old. Certain drugs like enalapril, monteleukast and vaccination with diphtheria-tetanus toxin have been shown to precipitate the disease. It is clinically characterised by oral mucosal erosion and flaccid vesicles and bullae, which break easily to form erosions and crust. 'Nikolsky sign', elicited by applying tangential pressure on the skin surface resulting in separation of the epidermis indicates acantholysis (loss of coherence between epidermal cells). 'Asboe Hansen sign'

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is elicited by gently pushing the bullous content and noting the spread of the bulla into the apparently normal surrounding. Involvement of mucous membrane mainly the oral mucosa has been seen in almost all cases and usually precedes the cutaneous lesions. Ocular involvement in childhood PV have been reported. The characteristic ocular finding is conjunctivitis with hyperemia and mucoid discharge. Conjunctival blisters, erosions, and synechiae are rare and long-term sequel is uncommon.

The diagnosis is confirmed by demonstrating the acantholytic cells by doing a Tzanck smear and histopathologically by the presence of supra basal bulla with row of tomb stone appearance of the basal cells. Direct immune fluorescence will show intercellular deposits of IgG sometimes IgM, IgA in the epidermis.

PV should be differentiated from other immunobullous diseases, impetigo, and erythema multiforme.

Treatment: Child should be hospitalized and the treatment should be tailored for each child depending on age, drug contraindication, side effects, area involved and disease activity. Corticosteroids are the cornerstone of treatment at an initial dose in range of 2-3 mg/kg/day with slow tapering to 0.5 to 0.8mg/kg/day in approximately 2 weeks following alternate day schedule for further reduction. Methylprednisolone pulse (1 gm/day I.V. for 5 days) and dexamethasone pulse (136 mg of dexamethasone/day I.V. for 3 consecutive days) has also been used successfully in severe cases.

Azathioprine, cyclosporine, methotrexate, gold can be used in selected children. Other useful drugs include dapsone, rituximab and plasmapheresis.

Pemphigus foliaceus (PF)

Childhood PF is slightly more common in boys with M: F ratio of 1.33:1 but gender does not correlate with age at presentation The trigger factors are sunlight, drugs, bacterial infection, cytomegalovirus infection and otitis media. The face, scalp or both are the sites most commonly involved initially followed by trunk or upper extremities. Lesions appear like scaly papules or very superficial short-lived vesicles that rupture soon to leave scaling. When the disease worsens, it leads to erythroderma with a mousy odor. Because of the chronicity of the disease, there may be stunting of growth. Mucus membrane involvement is not a feature of PF.

This should be differentiated from other causes of erythroderma like atopic dermatitis, seborrheic dermatitis, pityriasis rubra pilaris and staphylococcal scalded skin syndrome

The diagnosis is confirmed by demonstrating the acantholytic cells by doing a Tzanck smear and by the presence of sub corneal cleavage histologically. Direct immune fluorescence will show intercellular deposits of IgG in the epidermis.

Treatment: Systemic steroids alone are sufficient in most instances. Other therapies include hydroxychloroquine, erythromycin, chloroquine, methotrexate, sulfapyridine and azathioprine.

Paraneoplastic pemphigus (PNP)

PNP is characterised by features of pemphigus vulgaris having polymorphous lesions and severe intractable mucositis. Respiratory injury may lead to bronchiolitis obliterans and prominent lichenoid mucositis or dermatitis. Although adult patients are more commonly affected there has been an increasing number of individual reports in the child and adolescent population. PNP affects children and adolescent aged 8-18 years with no gender preference Unlike adult cases where NHL is the most commonly associated neoplasm, in children striking association with Castleman's disease is seen.

Histology may either show, lichenoid infiltrate with cell necrosis or intraepithelial acantholysis or combination of both. Direct immune fluorescence will show intercellular deposits of IgG and C3 in the epidermis and the sub epidermal region.

Treatment: None of the immunosuppressives is found to consistently work. Management of the underlying malignancy may remit the disease.

Sub epidermal immune bullous disorders

Sub epidermal immunobullous diseases are uncommon in childhood. In contrast to adults, the most commonly seen is IgA-mediated chronic bullous disease of childhood (CBDC), while IgG-mediated bullous pemphigoid (BP), bullous SLE and epidermolysis bullosa acquisita (EBA) are rare.

Chronic bullous dermatosis of childhood (CBDC)^{2,3}

CBDC is a distinctive sub epidermal blistering disorder of child hood . The eruption usually begins before the age of 6 years and is typically described as self-limiting, clearing within a few months or years. The disease often commences with urticated, annular to targetoid lesions with the subsequent development of the classic "cluster of jewels" lesions of grouped small blisters around the edge of an erythematous annular lesion. Milia and scarring occur in

the healing phase. Genital mucosal lesions may heal with scarring. The lesions characteristically involve the perioral area, lower trunk, inner thighs and genitalia. The classical finding is the "string of pearls" appearance. Direct immunofluorescence shows a linear band of IgA at the dermoepidermal junction (DEJ). Chronic bullous dermatosis of childhood has many clinical and immuno pathological similarities with adult linear IgA disease (LAD) and it has been suggested that they represent different manifestations of the same disease.

Treatment: Dapsone is considered to be the drug of choice for CBDC. Combination with cimetidine increases its tolerance and reduces hemato-toxicity by inhibiting the formation of hydroxylamine metabolites of dapsone. Cyclosporine and thalidomide are found to be useful in recalcitrant cases.

Childhood bullous pemphigoid (BP)^{4,5}

The clinical and immuno pathologic features of children with acquired sub epidermal blistering disorders show

considerable overlap, and their classification frequently requires characterization of the targeted antigens.

The following criteria should be fulfilled for a diagnosis of childhood BP:

1. 18 years of age or younger, with the clinical appearance of tense bullae and the histopathologic finding of sub epidermal bulla formation with a variable amount of eosinophils.
2. Direct IF showing linear deposition of IgG or C3 as the major immunoreactant at the basement membrane zone (BMZ) or a positive indirect IF demonstrating circulating IgG auto antibodies that were reactive with the BMZ.

BP rarely, occurs in children. It has been reported following solid organ transplant. The clinical features of BP include widespread tense blisters, arising on inflamed or normal appearing skin, that are predominantly located on flexural areas. In childhood-onset bullous pemphigoid associated with vaccination, the bullous lesions

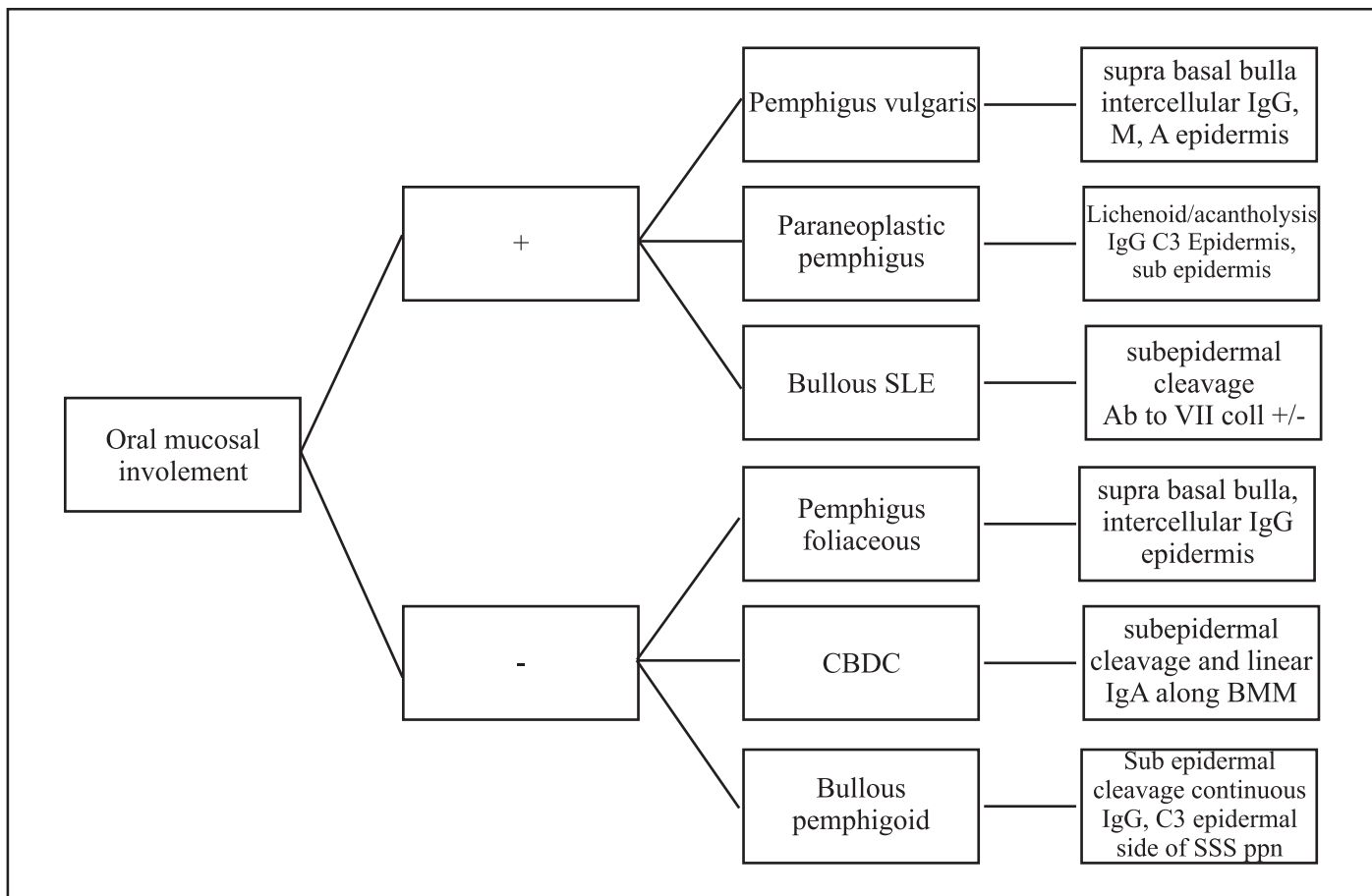


Fig.1.Simple algorithmic approach to a child with immune bullous disease

predominantly affect the palms, soles and face. In infants the blisters tend to occur frequently on the palms, soles and face, affecting the genital areas rarely; 60% of these infant patients have generalized blisters.

Sometimes it may be difficult to differentiate childhood BP from pompholyx and other sub epidermal diseases of childhood, including linear IgA bullous dermatosis and Epidermolysis bullosa aquisita.

Histologically the lesion shows sub epidermal bulla with eosinophils. Diagnostic features of BP include linear deposition of IgG, A, M and C3 along the dermal-epidermal junction on direct immunofluorescence (IF).

Treatment: Prednisolone 1-2 mg/kg daily. Other treatments with reported benefit are potent topical steroids alone, erythromycin as monotherapy or with nicotinamide, as well as sulfapyridine and dapsone, both as sole treatment, or with prednisolone, mycophenolate mofetil (MMF), rituximab and subcutaneous omalizumab are found to be useful.

Bullous systemic lupus erythematosus⁶⁻¹⁰

Bullous SLE usually presents with generalized eruption of tense vesicles and bullae with a non-inflamed base. By definition, all patients should satisfy the American Rheumatological Association criteria for diagnosis of SLE. Bullous eruption can be an initial manifestation of SLE.

The biopsy findings may mimic dermatitis herpetiformis with sub epidermal separation and neutrophil papillary micro abscesses. Basal cell vacuolation, Civatte bodies and dermal vasculitis are not seen in all affected children with SLE unlike in adult. Bullous SLE is divided into types I and II on the basis of the presence or absence of antibodies to type VII collagen. Failure to demonstrate these antibodies permits classification as type II BSLE. The significance of this sub classification is unclear as clinical differences between the two types are not apparent.

Treatment: Dapsone is the drug of choice in bullous SLE. Prednisolone, methotrexate, azathioprine and mycophenolate mofetil, are other drugs that are useful in the treatment of BSLE. Rituximab was found to be useful in the treatment of bullous SLE.

Points to Remember

- *Immunobullous disorders in children, though rare are not infrequent.*
- *High index of suspicion will help early diagnosis*

and management. Steroids form the main stay in pemphigus group of diseases with or without another immune suppressant.

- *Pulse therapy of steroid when administered carefully gives better results with lesser side effects. Pemphigoid has a relatively better prognosis when compared to pemphigus and hence can be treated with steroid alone.*
- *Dapsone is the drug of choice in CBDC and bullous SLE. Other drugs like IV Ig and biologic agents should be used only in selected cases where the other drugs fail.*

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RADIOLOGY

NASAL MASSES

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We have seen sinonasal polyposis as a cause of nasal masses. We will now see a few more. Nasal masses are rare. Some congenital masses that we see are dermoids, gliomas and encephaloceles all of which may have intracranial connections. Therefore all nasal masses in children should be evaluated as congenital masses with potential intracranial connections because a hasty biopsy can lead to meningitis or CSF leak.

Abnormalities of development result in the three kinds of masses mentioned above. Dermoids are commonly midline cystic masses. During embryological development, if the skin maintains attachment to underlying deeper tissues like the nasal capsule or dura some epithelial elements get pulled under the surface and dermoid forms. Dermoid cysts can get infected, form an abscess and lead to meningitis. Gliomas develop from extracranial rests of glial tissue. They are possibly encephaloceles which have lost connection with CSF. They are usually extra nasal but a third of them can be intranasal as the glioma in Fig.1. Fig.1 is an MRI showing a midline intranasal mass that is isointense to brain. There is no demonstrable stalk extending intracranially. Dural connection is more likely with the intranasal type. There is no connection with the subarachnoid space and hence they do not enlarge on crying or straining. Nasal gliomas are best resected early to avoid complications including nasal deformity. Encephaloceles are extracranial herniation of meninges with or without brain. The usual location is the occiput. The basal encephalocele is seen as an intranasal mass or a mass in the nasopharynx

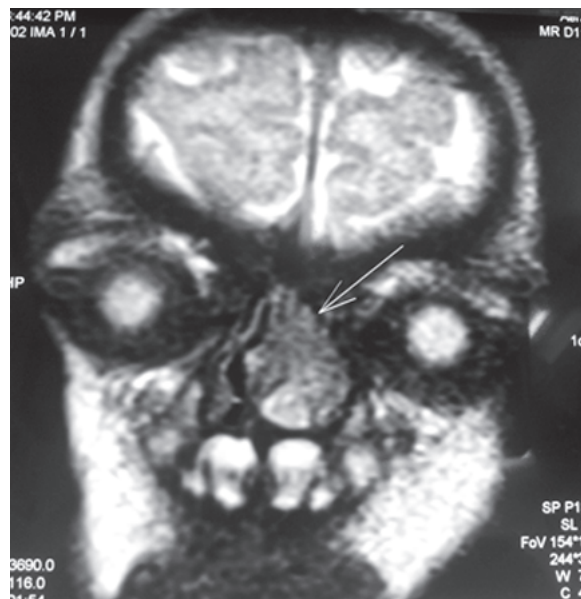


Fig.1. Midline nasal mass – Nasal glioma

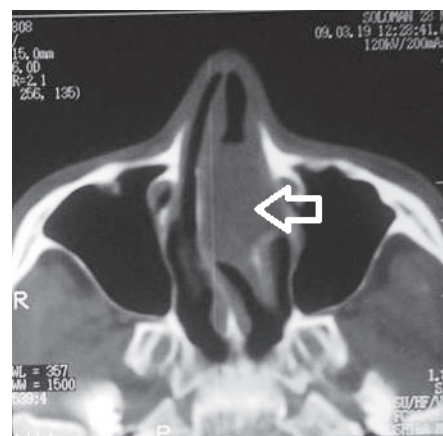


Fig.2. Transethmoidal encephalocele

or orbit. Encephaloceles enlarge on crying due to the intracranial communication, in contrast to nasal gliomas. Imaging is absolutely essential to determine the origin of a nasal mass. Fig.2 shows what looks like an innocuous polyp but further sections show the nasal mass coming down from inside the cranium through a gap in the cribriform plate and extending inferiorly medial to the turbinates Fig.3. This is a transethmoidal encephalocele. Basal encephaloceles can remain hidden clinically for many years. The transsphenoidal

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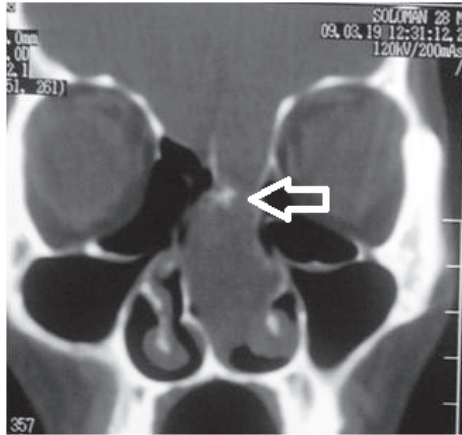


Fig.3. Transethmoidal encephalocele. Note the gap in the cribriform plate (arrow)



Fig.6. Rhinolith

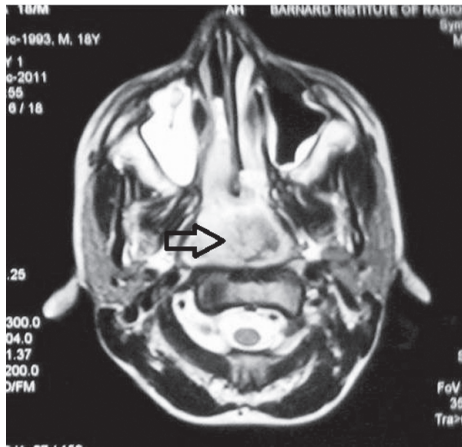


Fig.4. Juvenile nasopharyngeal angiofibroma.

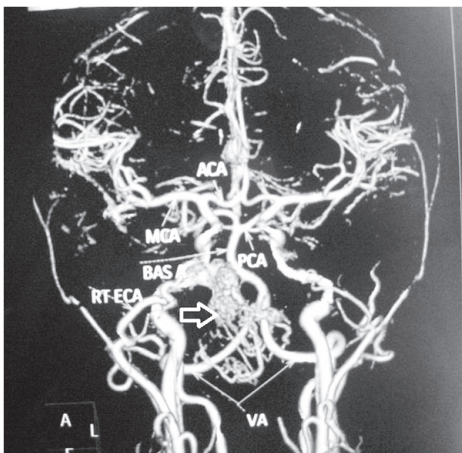


Fig.5. MRA same patient as Fig 4.

and sphenothmoidal basal encephaloceles present in the nasopharynx. CT is useful for delineating the bony gap while MRI is useful for extent of soft tissue herniation.

The juvenile nasopharyngeal angiofibroma (JNA) is another cause for nasal mass. It is a benign highly vascular mass that occurs almost exclusively in male adolescents. Although benign it is locally very aggressive and has a tendency to recur after excision. It arises from the roof of the nasopharynx in the region of the sphenopalatine foramen. From here these masses extend into the cavity of the nasopharynx and then into the pterygopalatine and infratemporal fossa. They classically cause bowing of the posterior wall of the maxillary sinus. The skull base can also be eroded. Clinically they present with a nasopharyngeal mass and epistaxis. CT and MRI will confirm the diagnosis and map the extent of tumour. CT brings out bony erosion while MRI shows the soft tissue extent. The pattern of mass in the nasal cavity and pterygopalatine fossa with bony erosion is diagnostic for JNA and there is no necessity for a dangerous biopsy. On MRI there maybe flow voids. There is marked enhancement following gadolinium administration reflecting the highly vascular nature of the mass. Complete excision of the mass is the goal of treatment and imaging will help in planning the approach for a thorough removal. Fig.4 shows a hyper intense mass filling the nasopharynx. The mass has obstructed the meatus of the right maxillary sinus resulting in hyper intense pent up secretions Fig.5 is an MR angiogram showing a number of branches from the right maxillary artery and some branches from the right internal carotid artery supplying the JNA.

One unusual nasal mass is the rhinolith. The rhinolith consists of a nidus of inspissated mucus, clot or even a small foreign body on which is deposited carbonates and phosphates of calcium that is present in nasal secretions. This goes on for many years till it causes nasal obstruction. Fig.6 is a rhinolith where you can see a lucent area around

a white nidus in the centre and opaque concretions in the periphery.

What is evident clinically as nasal masses may be just the tip of the iceberg. Imaging is very important to determine the nature and internal extent of the mass.

CLIPPINGS

Kizilca O, Siraneci R, Yilmaz A, Hatipoglu N, Ozturk E, Kiyak A, Ozkok D. Risk Factors for Community-Acquired Urinary Tract Infection with ESBL-Producing Bacteria in Children. Pediatrics International. 08/17/2012.

Recognition of the risk factors for extended-spectrum beta-lactamases [ESBL]-producing bacteria may be helpful to determine new policies in management of urinary tract infection. These results suggest that recurrence of UTI should be prevented especially in the first year of life and prophylactic use of cephalosporins should be avoided. Age below 1 year, high recurrence rate of UTI, long duration of prophylaxis, using cephalosporins for prophylaxis, having been hospitalized in the past 3 months and clean intermittent catheterization were found to be significant risk factors for ESBL producing bacteria.

Paul R, Neuman MI, Monuteaux MC, Melendez E. Adherence to PALS Sepsis Guidelines and Hospital Length of Stay (LOS). Pediatrics, 08/13/2012.

Overall adherence to Pediatric Advanced Life Support sepsis guidelines was low; however, when patients were managed within the guideline's recommendations, patients had significantly shorter duration of hospitalization. Patients who received 60 mL/kg of intravenous fluids within 60 minutes had a 57% shorter hospital LOS ($P = 0.039$) than children who did not. Complete bundle adherence resulted in a 57% shorter hospital LOS ($P = 0.009$).

NEWS AND NOTES

Twelfth ICMR Course on Medical Genetics and Genetics and Genetic Counseling, Uttar Pradesh

Date: July 29th – August 10th, 2013

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CASE STUDY

LYMPHADENOPATHY - A DIAGNOSTIC CHALLENGE

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Abstract: *Chronic lymphadenopathy in children can sometimes be diagnostic dilemma. A seven year old female child presented with isolated left sided posterior cervical lymphadenopathy. Investigations for tuberculosis and lymphoma were negative. Histopathology helped to arrive at the rare diagnosis of Kikuchi -Fujimoto disease.*

Keywords: *Lymphadenopathy, Kikuchi Fujimoto disease, Children*

Case Report

A seven year old girl presented with continuous high grade fever and cervical lymphadenopathy of four weeks duration. Since symptoms and lymphadenopathy persisted in spite of antibiotics, child was referred to rule out tuberculosis. There was no history of contact with tuberculosis.

On examination, she was pale and febrile. A single lymph node of about five centimetres was palpable in the left posterior cervical area. The node was non tender but firm in consistency. No other group of lymph nodes were palpable. No foci for infection were identifiable. Liver was palpable two cm below the right costal margin and there was no other organomegaly. Examination of other systems was within normal limits.

Blood smear showed microcytic, hypochromic anemia, normal platelet count and the hemoglobin was 9.5 gm%. The total count was 4,600 / μ L with lymphocyte predominance. Chest skiagram was normal and fever workup including blood and urine cultures were negative. Ultrasound abdomen showed mild hepatomegaly.

Fine needle aspiration cytology (FNAC) from the node was suggestive of nonspecific lymphadenitis. During hospital stay, the child developed nonpruritic erythematous maculopapular rashes all over the body suggestive of erythema multiforme.

Hematologist suggested excision biopsy and immunohistochemistry to rule out lymphoma. The lymph node was excised in toto and histopathology revealed total replacement of lymph node architecture with focal areas of necrosis, surrounded by lymphocytes, eosinophils, cluster of histiocytes, karyorrhectic debris and immunoblast like cells with mononucleate forms, prominent foamy histiocytes and absence of neutrophilic infiltration- the picture suggestive of necrotizing type of Kikuchi-Fujimoto disease (KFD).¹

As the fever persisted and the association of KFD with systemic lupus erythematosus is well recognised, antinuclear antibody (ANA) and dsDNA were performed. ANA was 3+ positive and dsDNA was negative. A diagnosis of Kikuchi-Fujimoto disease associated lupus like syndrome was arrived at and she was started on prednisolone and hydroxychloroquine. The child became afebrile within a week.

Discussion

Kikuchi-Fujimoto disease or histiocytic necrotizing lymphadenitis is an uncommon, idiopathic, generally self-limited cause of lymphadenitis.^{2,3} Although several causes like *Yersinia enterocolitica* and *Toxoplasma gondii* and viruses including Epstein-Barr virus (EBV), herpes virus⁶, parainfluenza virus and cytomegalovirus are implicated, none have been strongly proved to be the etiological agent.⁴ An autoimmune mechanism has also been proposed because KFD is seen in conjunction with systemic lupus erythematosus (SLE).⁵

The diagnosis of KFD is challenging because of lack of specific signs, symptoms and serological markers. The clinical presentation of KFD is very similar to lymphoma, tuberculosis and systemic lupus erythematosus. This patient presented with fever, cervical lymphadenopathy and rash, but other manifestations like maxillary and mesenteric lymphadenopathy, splenomegaly, parotid gland

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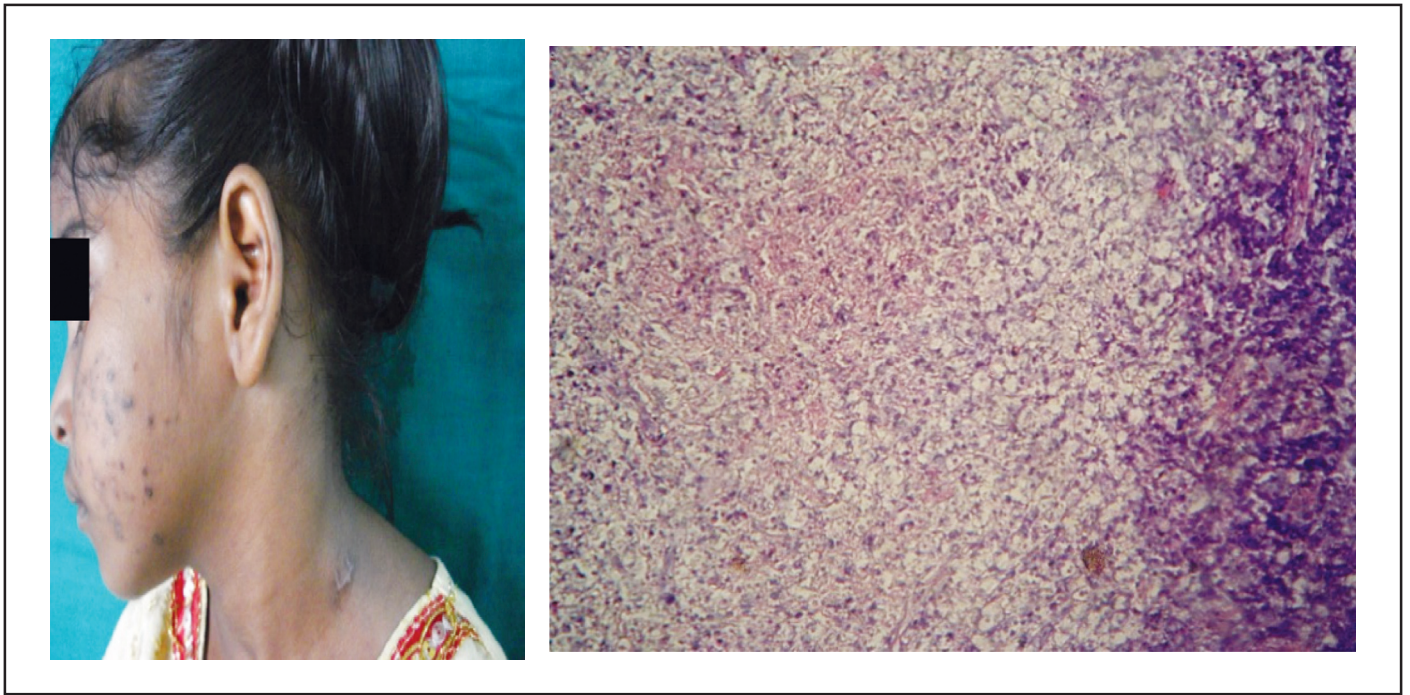


Fig.1. Resolved maculopapular rashes over the face, site of excision biopsy.

Fig.2. Histopathology showing areas of necrosis, karyorrhectic debris, foamy histiocytes, absence of neutrophils.

enlargement, arthralgias, myalgias, aseptic meningitis, bone marrow hemophagocytosis and interstitial lung disease were observed in addition by other authors.^{6,7,8,9} In their comprehensive review on KFD, Bosch X et al have cited various other unusual manifestations of KFD like thyroiditis, carcinoma and diffuse large B-cell lymphoma. Cases of KFD have been associated with autoimmune conditions other than SLE and presented as pyrexia of unknown origin.⁶

The erythematous papules observed in this child could have been part of the cutaneous spectrum of KFD which includes facial rash, exudative erythema, erythematous papules, vasculitis and nodules which are observed in 40% of cases.¹⁰ Though the lymph node was completely excised in our case the usual course is self limiting and resolves over several weeks to 6 months.¹¹

When FNAC is not contributory, biopsy and HPE should always be done as it helps to arrive at a precise diagnosis and is considered the gold standard.¹² Immunohistochemistry (IHC) of the histiocytes in KFD expresses CD11b, CD11c, CD14, CD68, lysozyme and MPO, a unique feature seen in KFD.⁹ Ki MIP, a marker of plasmacytoid monocytes is useful in differentiating KFD from lymphoma.¹³

Though some authors recommend corticosteroids in severe extra nodal or generalized disease, KFD is a self-

limiting disorder and resolves within one to four months and does not require specific management.^{11, 14} Positive ANA or with systemic symptoms warrants steroids as in our case.^{15,16} Children diagnosed as KFD should be evaluated for SLE as the former is considered a “forme fruste” of SLE.¹⁷ This has been demonstrated in our case where ANA was positive.

Empirical antituberculous drugs should be discouraged even in cases of isolated lymphadenopathy in a TB endemic country like ours, and an attempt at histological or bacteriological proof for disease should be made in all cases, as they can reveal completely different diagnoses as it was in this case.¹⁸

Conclusion

Though unilateral posterior cervical lymphadenopathy may be caused by several conditions like tuberculosis and lymphoma, at times disorders like KFD should also be considered in atypical presentations, where biopsy helps to clinch the diagnosis.

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Nabulsi M, Hani A, Karam M. Impact of C-reactive protein test results on evidence-based decision-making in cases of bacterial infection. BMC Pediatrics, 09/17/2012.

The routine ordering of C-reactive protein (CRP) for children with infections is based on weak evidence. The impact of the CRP test results on decision-making is rather small and CRP ordering may contribute to unnecessary health care expenditures. Better quality research is needed to definitively determine the diagnostic accuracy of CRP levels in children with infections.

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CASE STUDY

GANGLIONEUROBLASTOMA PRESENTING AS VERNER MORRISON SYNDROME

***Latha Vishwanathan**

****Anita Tarigopula**

*****Sripathi V**

******Harish S Kumar**

*******Malathi Sathiyasekaran**

Abstract: *Ganglioneuroblastoma secreting vasoactive intestinal polypeptide and manifesting as a triad of watery diarrhea, hypokalemia and achlorhydria (WDHA) or Verner Morrison syndrome (VMS) is a rare entity. We report a 4 year old boy with recurrent watery diarrhea and hypokalemia diagnosed as VIP secreting ganglioneuroblastoma with complete resolution of symptoms following surgery and on regular follow up.*

Keywords: *Ganglioneuroblastoma, VIP, Watery diarrhea, Verner Morrison syndrome.*

Verner Morrison Syndrome (VMS) is a rare neuroendocrine tumor with an incidence of 0.05 to 0.2 cases per million adults. It is usually due to Vasoactive Intestinal Peptide (VIP) secreting tumor of the pancreas but may arise from adrenals in children. The incidence in pediatric population is not known and is rarely reported from India¹. Approximately 50% of VMS have been identified as VIP secreting ganglioneuroblastomas. This case report highlights the necessity to consider VIPoma in any child with recurrent watery diarrhea and persistent hypokalemia since it may be amenable to curative surgery.

Case Report

A 4 year old boy presented with watery diarrhea and

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failure to thrive since 2 years of age. There was no history of fever, pedal edema, flushing, anasarca, GI bleed, skin rash, wheeze or arthralgia. He was mildly dehydrated without pallor or significant lymphadenopathy. His weight and height were above the 50th centile. Blood pressure was 90/60 mm of Hg. System examination was normal except for mild abdominal distension. CBC, urine, stool, liver biochemistry, renal function tests, cholesterol and X ray chest were normal. Serum sodium and chloride were normal, bicarbonate was 16mEq/L but potassium was persistently low ranging from 1.5 to 2.9 mEq/L despite repeated corrections. A possibility of VMS was considered; however the initial ultrasound abdomen was normal. Since immunoglobulin and lipid profile, HIV, tissue transglutaminase antibody and upper GI endoscopy with duodenal biopsy were all normal, a repeat ultrasound was done which revealed a mass near the tail of pancreas which had not been identified earlier. CECT abdomen delineated a well defined, oval retroperitoneal mass with multiple calcifications compressing on the anterior aspect of the left kidney without enhancement of the adjacent lymph nodes (Fig.1). Serum gastrin, urinary catecholamines and vanillyl mandelic acid (VMA) were normal. Analysis of gastric juice showed achlorhydria. The above investigations were diagnostic of Verner Morrison Syndrome. Surgically an encapsulated mass with multiple calcifications free from the pancreas, adrenal and left kidney weighing 170 grams, 8.0 x 6.0 x 4.5cms and without involvement of the adjacent lymph nodes was resected (Fig.2). Histopathological

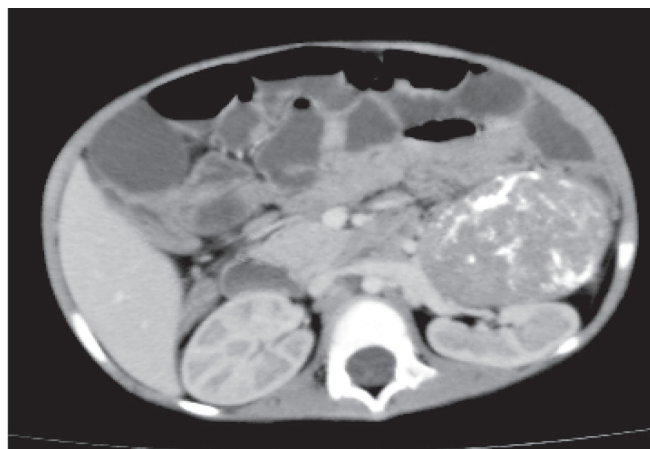


Fig.1. CECT showing mass with calcifications



Fig.2. Resected tumor measuring 8.0cm x 6.0cm x 4.5cm and weighing 170 Grams.

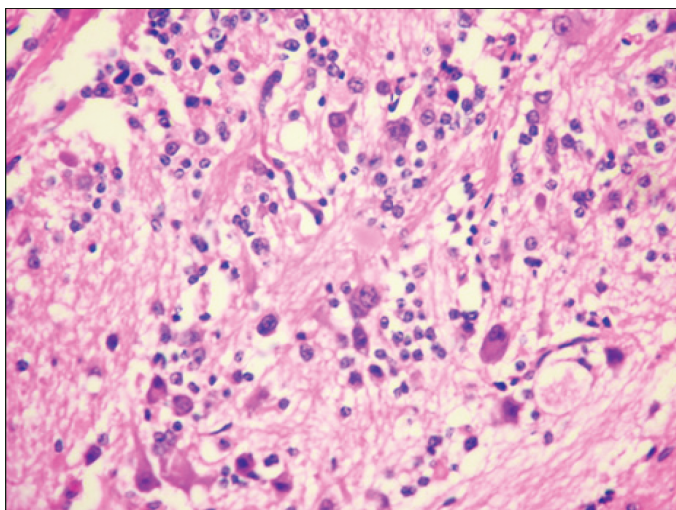


Fig.3. Ganglioneuroblastoma showing sheets of neoplastic cells composed of immature and mature ganglion cells in high power field.

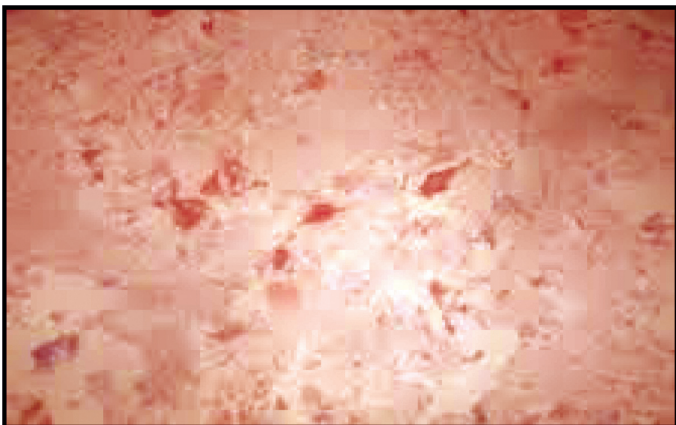


Fig.4 VIP stain positivity.

examination (HPE) revealed ganglioneuroblastoma stroma poor type (International Neuroblastoma Pathology classification) (Fig.3) and mitosis karyorhexis index of <1% indicating a favorable histology. Immunohistochemistry showed chromogranin, S100, synaptophysin and intense VIP stain positivity (Fig.4). Adjuvant chemotherapy was not suggested by the concerned oncologist. Child had complete resolution of clinical and biochemical findings following resection and is on regular follow up for the past 1year.

Discussion

Verner and Morrison in 1958 described a syndrome of watery diarrhea, hypokalemia, and achlorhydria (WDHA) associated with non-insulin secreting tumors of the pancreatic islets.² In 1957 Priest and Alexander had reported a similar syndrome of watery diarrhea, hypokalemia and hypochlorhydria in an adult with pancreatic endocrine tumor. Swift et al in 1975³ were the first to report VIP secreting ganglioneuroma in a child. VIPomas usually (90%) arise in the pancreas in adults whereas in children and adolescents, ganglioneuromas, ganglioneuroblastomas, neurofibromas or adrenal tumors may present as VIPoma. The tumors may be situated in the retroperitoneum, thorax, paraspinous region, neck, intestine or adrenals. VIP is a biologically active 28 amino acid polypeptide and causes intestinal secretion by stimulation of adenylate cyclase. Splanchnic and systemic vasodilatation and inhibition of both pentagastrin and histamine stimulated gastric acid secretion by VIP may explain the other clinical features.

VMS usually presents around 2 months to 11years of age. The majority of these tumors (55%) are ganglioneuroblastoma and present with intractable diarrhea, hypokalemia and metabolic acidosis as in this case⁴. The common differential diagnosis in Indian children presenting with watery diarrhea, hypokalemia and failure to thrive include celiac disease, HIV and tuberculosis. If a mass is detected other neuroendocrine tumors may be considered. Ultrasound (USG), computed tomography (CT) and magnetic resonance imaging are modalities that are useful in diagnosis. The sensitivity of USG and CT is 50-80% in tumors more than 3cms.⁵ Endoscopic ultrasound increases the diagnostic yield when the lesions are small.⁶ In this child the mass was not identified in the initial ultrasound probably due to the dilated bowel loops. Abdominal distension and colonic dilatation is an effect of VIP and may be the reason for this discrepancy.⁷ Serum VIP estimation helps in diagnosis but was not estimated in this child due to non availability. However the intense positive VIP staining of the tumor in addition to the clinical and biochemical features confirmed the diagnosis.⁸

VIP expression in ganglioneuromas and neuroblastomas correlating with a more favorable prognosis is controversial.⁹ In adults somatostatin scintigraphy using various somatostatin analogues which bind with the high affinity receptors overexpressed in these tumors have been included in the diagnostic armamentarium.¹⁰

The initial treatment of VMS is directed toward correcting fluid and dyselectrolytemia. Somatostatin analogues such as octreotide acetate may temporarily decrease the diarrhea and suppress VIP levels. However surgery offers the definitive cure in cases where tumor is resectable.¹⁰ When the tumor is unresectable combo chemotherapy with doxorubicin and streptozocin may be administered¹⁰. In this child the tumor was encapsulated and free from the adjacent organs and hence could be resected in toto. Adjuvant chemotherapy was not initiated since the HPE and mitosis karyorrhexis index favored a good prognosis. The child was reviewed one year later and had no recurrence of symptoms.

Verner Morrison Syndrome though rare should be considered in any child with watery diarrhea and persistent hypokalemia since the lesion may be amenable to definitive surgery. Though the diagnosis may be delayed despite initial biochemical and radiological investigations, a diligent search may be rewarding.

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CLIPPINGS

Krogh C, Biggar RJ, Fischer TK, Lindholm M, Wohlfahrt J, Melbye M. Bottle-feeding and the Risk of Pyloric Stenosis. Pediatrics Oct 2012

Bottle-fed infants experienced a 4.6-fold higher risk of pyloric stenosis (PS) compared with infants who were not bottle-fed. The result adds to the evidence supporting the advantage of exclusive breastfeeding in the first months after birth. The increased risk of PS among bottle-fed infants was observed even after 30 days since first exposure to bottle-feeding and did not vary with age at first exposure to bottle-feeding.

Kanemura H, Sano F, Mizorogi S, Aoyagi K, Sugita K, Aihara M. Duration of recognized fever in febrile seizure predicts later development of epilepsy. Pediatr International 03/23/2012.

The duration of recognized fever appears to provide useful information about the risk for the later development of epilepsy. Either an unusually short or long duration of recognized fever prior to the initial FS was associated with an increased risk of unprovoked seizures. Either an unusually short or long duration of recognized fever prior to the initial FS was associated with an increased risk of unprovoked seizures.



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09.20-09.30 am	Discussion	
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12.35-12.50 pm	● Noisy breathing	Dr.L.Subramaniam
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