IAP GOA E-Bulletin

BULLETIN *October 2019*

Activities from

July 2019 to September 2019

Issue 8

GOA STATE CHAPTER

For Private Circulation

Table of Contents:

Editors Note...

Greetings to all the IAP Members,

With the rains having a retreating trend and we paediatricians have a little space and time to breathe, we bring to you our next issue of e-bulletin IAP Goa State Chapter.

The last 3 months have made us realize one thing…. Nature is the supreme power … No wealth no human innovation can defeat it. Incessant rains wrecked havoc in atleast nine states including entire Peninsular India. Natural calamities not only cause a huge burden of deaths but also lead to ongoing morbidity. The new bugs that arise are not only aggressive and resistant but also difficult to identify and hence control. They also pose a threat of epidemics and hence a watchful eye is required in order to identify and break the epidemic link.

In the 8th issue of IAP Goa State Chapter e-bulletin we have discussed topics such as traumatic brain injury, Burkitt's lymphoma, thalassemia , neuroblastoma and thyroid hormone transport proteins, which greatly benefit our day to day practice and is of great concern in the health care of children.

A special mention must be made about Breastfeeding week which was celebrated in many Government n Private hospitals with a lot of enthusiasm. An effort truly well done and well appreciated.

I thank immensely all the members who have taken time from their busy schedule and contributed to the learning process.

In the words of Jillian Michael -"It's not about being perfect. It's about effort and when you bring that effort every single day, that's where the transformation happens, that's how change occurs"

Warm regards

Dr. Priyanka Dhakankar

CASE REPORT: An interesting case of Burkitt's lymphoma

Dr Roshen Mascarenhas
DCH, DNB
(Ex-Senior Resident)
Kashiben Gordhandas Patel Children's Hospital, Baroda.

INTRODUCTION

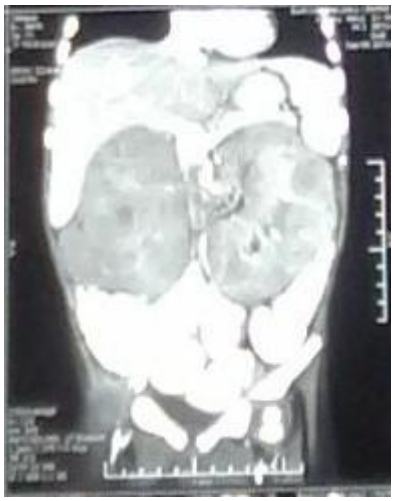
Burkitt's lymphoma is a very aggressive and most frequent subtype of non-Hodgkin lymphoma in childhood and accounts for approximately 34% of these cases. It is composed of small, non-cleaved, diffuse, undifferentiated malignant cells of lymphoid origin. Burkitt's lymphoma was first described by Dennis Burkitt in 1958 while he was studying this tumour in Africa where it was endemic. It is characterized by the deregulation and translocation of the C-MYC gene from chromosome 8. It is the most rapidly growing tumour in children with a doubling time of approximately 24 hours, hence, prompt recognition and initiation of therapy are essential.

CASE REPORT

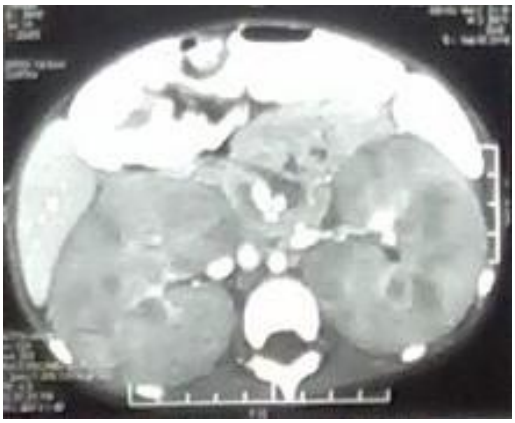
A three year old male child born out of non-consanguineous marriage presented with history of abdominal distension, swelling of face and fever on and off for 15 days. Examination revealed a well-nourished child, irritable with toxic appearance and normal vitals. Significant swelling in the bilateral maxillary region was noted. Swelling was hard, non-tender with no local rise of temperature or skin changes. Systemic examination revealed diffuse abdominal distension with bilateral palpable masses in the flank which were firm, ballotable and non-tender. There was no evidence of hepatosplenomegaly or lymphadenopathy. Other systemic examination was unremarkable.

Investigations revealed anaemia with normal counts. Serum RFTs, electrolytes, LFTs, chest X-ray and Urine microscopy were normal. Serum phosphorous (7.9 mg/dl), serum uric acid (11mg/dl) and serum LDH (3186 U/l) were high. Abdominal ultrasound revealed bulky kidneys with possible intussusception. Provisional diagnosis considered at this point was either a neuroblastoma or a lymphoma. Child was started on hydration and urine alkalinising measures in view of tumour lysis. CT abdomen revealed bilateral nephromegaly, mesentric edema, thickening & omental thickening in iliac fossa region. Intussusception at colo-colic and transverse 1/3rd of colon was present. These features were suggestive of lymphoma of colon and kidneys. CT face revealed large lobulated hypodense,

heterogenous soft tissue density in bilateral maxillary sinuses eroding the walls of maxillary antrum and extending into orbits and oral cavity. Biopsy of the right maxillary swelling showed evidence of sheets of lymphoid cells and necrotic bony fragments. Tumor cells expressed CD20 on immunohistochemistry. The diagnosis of Burkitt's lymphoma was confirmed and chemotherapy was initiated with injection Cyclophosphamide, Vincristine and steroids. Hydration, phosphate binders and oral allopurinol was continued. By the day 5 the general condition of the child improved, abdominal and facial swelling decreased. Phosphorous and uric acid levels normalised. Bone marrow and CSF analysis did not show any evidence of metastasis.

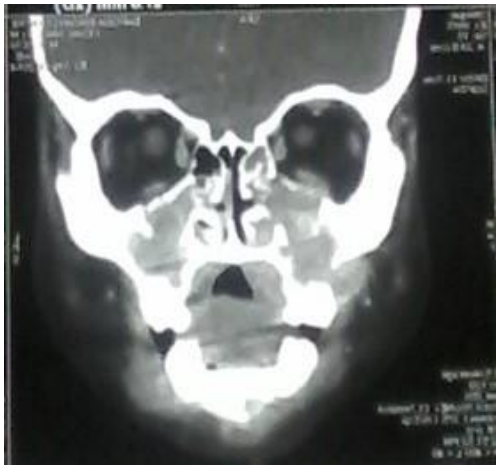


(Fig 1)



(Fig 2)

Coronal (fig 1) and axial (fig 2) view of CT abdomen showing bilateral nephromegaly, mesenteric edema, thickening & omental thickening.



(Fig 3)

CT face depicting heterogenous soft tissue density in bilateral maxillary sinuses eroding the walls (intraorbital extension, premaxillary extension, bilateral oral extension).

DISCUSSION

The WHO has classified Burkitt's lymphoma into three forms: endemic, sporadic, and immunodeficiency-associated types. The endemic form is commonly seen in males (M: F 2:1), mean age of presentation is between 3 to 8 years. It mainly affects the facial bones typically the maxilla and mandible and is usually linked to EBV virus. Sporadic subtype usually presents in children above 11 years and mainly affects the terminal ileum, the caecum and the intra-abdominal lymph nodes [(4)].They may present with an abdominal mass, intestinal obstruction and acute abdominal pain. Intussusception upon presentation can be present in up to 18% of the patients with primary abdominal Burkitt's lymphoma. Hematogenic dissemination is the most common cause for renal lymphoma. Adequate tissue is paramount to the diagnosis. Excisional biopsy of the lymph node or pleural fluid may be sampled for diagnosis. Histologically, Burkitt lymphoma comprises of sheets of medium-size lymphoid cells with scattered macrophages which have abundant pale cytoplasm. This creates a "starry sky" appearance. Immunohistochemical stains CD-19, CD-20, CD-22; CD-79a protein may be useful in diagnosis. Patients with any of the three clinical variants are at risk of spread to the central nervous system and bone marrow; hence bone marrow analysis and cerebrospinal fluid evaluation ought to be performed to assess the extent of involvement. Burkitt lymphoma is highly chemo sensitive and responds well to the treatment hence rapid cell lysis of the high tumour burden can lead to "tumour lysis syndrome" as noted in our patient during the course of treatment. Intensive chemotherapy has allowed 5 years disease free survival in early stages up to 90%. Age, LDH value, bone marrow and central nervous system involvement are the most frequently used prognostic factors.

KEY MESSAGES

- Burkitt´s lymphoma despite being very aggressive is potentially curable; therefore early diagnosis of this condition is essential.
- Tumor lysis syndrome is a common complication anticipated once chemotherapy is initiated.
- There can be overlap of presentations as seen in our case where features of both endemic and sporadic were present.

REFRENCES

1) Cadavid L, Sastoque JM, Gutiérrez C, et al. Primary osseous Burkitt lymphoma with nodal and intracardiac metastases in a child. Radiol Case Rep 2017; 12:185–90.

2) Molyneux EM, Rochford R, Griffin B, et al.Burkitt's lymphoma. Lancet 2012;379:1234–44.

3) Bonnet C, Janssens A, Wu KL, et al. BHS guidelines for the treatment of Burkitt's lymphoma. Belg J Hematol 2015;6:61–9.

4) WHO Press, Swerdlow SH, Campo E, Harris NL, et al. WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. Vol. 2. 2008;439.

5) Gupta H, Davidoff AM, Pui CH, et al. Clinical implications and surgical management of intussusception in pediatric patients with Burkitt lymphoma. J Pediatr Surg 2007;42:998–1001.

6) Patel PJ, Bahakim HM, Kolawole TM. Renal sonography in childhood lymphoma. Urol Int 1990;45:34–7.

Managing Traumatic Brain Injury: non-surgical management and the role of the paediatrician- a case based approach

Dr. Sumant Prabhudesai,
Consultant Paediatrician, Healthway hospital

"No brain injury is too mild to ignore or too severe to lose hope"

Introduction

Traumatic Brain Injury is a potentially life threatening condition often requiring aggressive and exhaustive management. Though "head trauma" intuitively appears to be a neurosurgeon's domain, in reality, it is often the paediatrician and intensivist that plays the larger role in the management of such patients. This is a case based discussion on the non-surgical aspects of managing children with severe traumatic brain injury.

Case presentation:

An 18 month old girl was brought to hospital after having fallen off a two-wheeler. She had lost consciousness immediately after the fall. What should the approach be to such a patient?

Discussion: Though this appears to be a case of head injury, children with vehicular trauma are likely to have multiple problems. All trauma patients should be assessed in the emergency bay. A systematic approach based on the PALS guidelines is important so as to not miss out important and life threatening problems.

The assessment usually begins with a quick "initial assessment" to ensure that the patient is conscious, is breathing and is adequately perfused, essentially ruling out a cardiac or respiratory arrest.

Subsequently, a "primary survey" is performed. This involves assessment of the airway, breathing, circulation, disability (neurology) and exposure. Any life threatening condition discovered during this stage needs to be addressed then and there before moving on to further examination.

In particular, children with severe traumatic brain injury are likely to be unable to maintain their airway and often need airway support such as jaw thrust,

oropharyngeal airway, bag and mask ventilation etc. until a definitive airway can be secured. It is important to remember that cervical spine injuries are more common in children with polytrauma than adults. C- spine injuries cannot be ruled out clinically in an unresponsive child. Cervical spine stabilisation (towels/ sand bags/ manual in-line stabilisation) should be done until c- spine injury can be ruled out.

Disability assessment in this situation involves level of consciousness (GCS or AVPU scale), pupils, tone and abnormal movements, decorticate or decerebrate posturing and XXX. A GCS or $\leq$ 8, unreactive pupils, unequal pupils and posturing are ominous signs and need to be addressed emergently. Similarly, bradycardia, hypertension and irregular respiration are signs if impending herniation and require emergency interventions. Hypotension is poorly tolerated by an injured brain and should be avoided.

A more detailed "secondary survey" can be done once major life-threatening problems have been identified and addressed. This survey involves a focussed clinical examination of the head and neck region, chest, abdomen, pelvis back and extremities.

Getting a CT head in this child is extremely important, but not more important than stabilising her.

Case continues:

On examination, the child had a patent airway and was hyperventilating. Her chest was clear, work of breathing was normal and SpO2 was 100% in air. Her heart rate was 160/min, pulses were well felt, extremities were cool, CRT 3 seconds and non-invasive BP 80/50 (64) mmHg. She had a GCS of 8 (E2 M3 V3). Her right pupil was 8 mm, not reacting to light, left pupil was 4 mm, reacting well. Her tone was normal and she had abnormal flexion to pain. How shall we proceed?

Comments: This child has "severe" Traumatic Brain Injury as her GCS is $\leq$ 8 and she has anisocoria. She is likely to have raised intracranial pressure (ICP) which if left uncontrolled can cause severe neuroparenchymal damage and result in poor neurological outcome including death.

Though she has a "patent" airway she is unlikely to be able to "protect" it and will require an emergency endotracheal intubation. In this situation, intubation is a high risk procedure and appropriate planning and preparation are vital. Until then, bag and mask ventilation can be safely performed.

Unilateral pupillary dilatation is a sign of temporal lobe (uncal) herniation. This is potentially reversible if acted upon quickly. Emergency measures for impending herniation include brief hyperventilation (in this case, B&MV) hyperosmolar therapy (hypertonic saline or mannitol if the BP is normal) and deep sedation.

She is also likely to be slightly hypovolemic and vasoconstricted and would benefit from a small fluid bolus especially since an intubation is being planned. She does not appear to have any chest or abdominal injuries.

Case continues:

Bag and mask ventilation was started with 100% oxygen sans head tilt-chin lift, using a jaw thrust only. Intravenous access could not be obtained as her extremities were cold. Intraosseus access was secured in both femurs (upper end). A normal saline bolus of 10 ml/kg was started in one and a bolus of 3% saline was started on the other. She was premedicated with lignocaine, induced with ketamine and fentanyl and paralysed with rocuronium. She was intubated with a 4.5 uncuffed endotracheal tube. Tube position was confirmed by auscultation and using capnography. A CT head was performed which showed no evidence of haemorrhage, midline shift or cerebral edema. There was no evidence of c- spine injury The scans were reviewed by the neurosurgeon and conservative management was planned. She was shifted to the PICU and mechanically ventilated.

Comments:

Rapid sequence induction plays an essential part in the intubation of children with possible raised ICP and should be performed even in a seemingly “obtunded” or unresponsive child. The very acts of laryngoscopy and intubation are sufficient to

cause a sharp spike in the already elevated ICP and result in herniation and even death. Lignocaine to some extent prevents such an ICP surge. Traditionally thiopentone has been used as an induction agent as it reduces cerebral metabolism and reduces the risk of excitotoxicity. However, hypotension is a common side effect, in this case, unacceptable. Recent studies show that ketamine is safe in raised ICP and may even be protective. Depolarising neuro-muscular blockers such as succinyl choline are contraindicated as they cause fasciculations which further raise the ICP. Non-depolarising blockers are routinely used.

The aim of a CT head here is to know the type of injury and whether neurosurgical intervention would be necessary. A neck CT can be performed simultaneously to rule out cervical spine injury.

The most common imaging abnormalities seen in severe TBI are extradural hematoma, subdural haemorrhage, intraparenchymal bleeds and an entity called "diffuse axonal injury" (DAI). A large number of children with severe TBI in fact have DAI as the predominant pathology. DAI occurs due to shearing forces during high velocity acceleration/deceleration injuries. CT findings may be non-specific and a normal scan does not rule out DAI. There is no role for primary surgical intervention here.

Case continues:

The child was shifted to PICU and mechanically ventilated on SIMV (PC) with a PEEP of 4 cm H2O and a targeted tidal volume of 6 ml/kg adjusted based on pCO2 and EtCO2. She was placed supine with the head in mid-position and elevated to 30^0. A right internal jugular central line was inserted and a right femoral arterial line was secured. A noradrenaline infusion was started targeting a mean arterial pressure (MAP) of 70. She was sedated with a midazolam and fentanyl infusion. A 3% saline infusion was started. IV fluids (5% DNS) were started aiming for a total fluid delivery of 80% of the calculated maintenance volume. Trophic feeds were started. A loading dose of levetiracetam was given.

Comments:

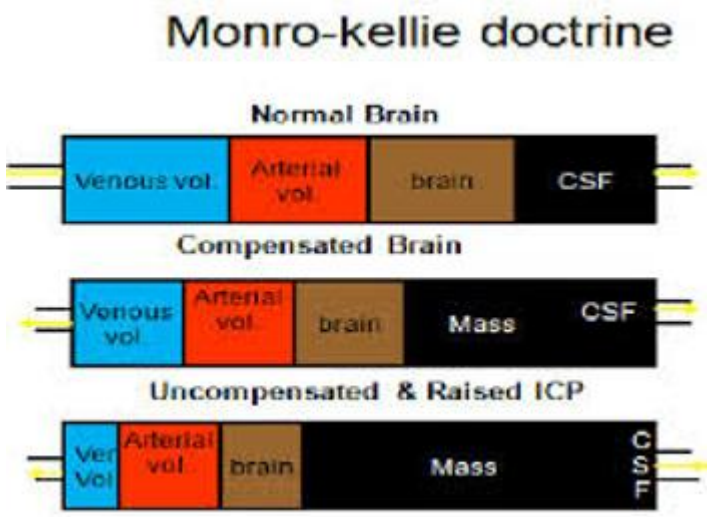
The aim of management in this child is now to prevent secondary brain injury. Primary brain injury (head trauma) as such cannot be reversed through interventions; the brain will recover over time provided normal homeostasis is maintained and secondary injury is prevented during this period.

Common entities that lead to secondary brain injury are hypotension, hypoxia, hypercarbia, fever, hypo- and hyperglycaemia. Of these, hypotension has been associated with the worst outcome. Therefore, maintaining adequate cerebral perfusion is the most important intervention here.

It is worth revisiting a few physiological principles to understand our management better.

Munro Kellie principle:

The Munro Kelly doctrine describes how a rise in the volume of one component of the intra-cranial contents results in a compensatory reduction in the volume of other components. An injured brain, like other injured organs tends to develop edema due to which there is a compensatory reduction in cerebral blood flow resulting in cerebral ischemia (*Figure 1*).



Intracranial pressure (ICP) :

ICP is normally < 10- 15 mm Hg and an ICP > 20 mm Hg is called intracranial hypertension. ICP is well known to rise in severe TBI particularly during the first 48- 72 hours. A high ICP over a prolonged duration in known to be associated with poor neurological outcome and interventions designed to lower the ICP are known to improve the outcome.

The only reliable clinical marker for suspecting a raised ICP in TBI is a low GCS. Clinical signs such as papilledema, Cushing's triad, posturing are fairly late signs and their absence should not be considered as evidence of its absence. Similarly, a normal CT scan does not rule out raised ICP. Currently, the only way to definitely know whether the ICP is raised or not is to measure it directly.

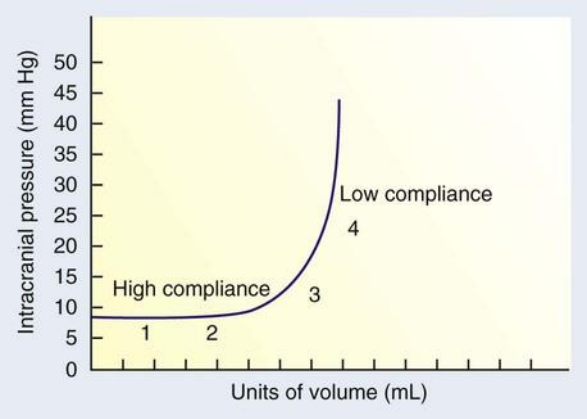
ICP can be measured through several different types of intracranial probes- epidural, subdural, intraparenchymal, intraventricular etc. Each has its own advantages and disadvantages but by and large, intraparenchymal probes are preferred globally. These probes allow continuous measurement of the ICP so that ICP-directed interventions can be effectively carried out.

A major limitation for the use of ICP monitoring is the availability of the equipment, availability of technical skill and costs. Currently, in India, ICP monitoring a done in a handful of tertiary care centres particularly in Tier 3 cities. Non-invasive ICP monitoring is currently being researched.

In the absence of ICP monitoring, it is safe to assume that the ICP is raised and treatment should be instituted accordingly.

Intracranial pressure-volume curve:

At normal cerebral/ CSF volumes, a rise in volume results in only a modest rise in ICP. However, at higher brain volumes (e.g. in cerebral edema), a slight increase in brain volume results in a dramatic rise in ICP (*Figure 2*).

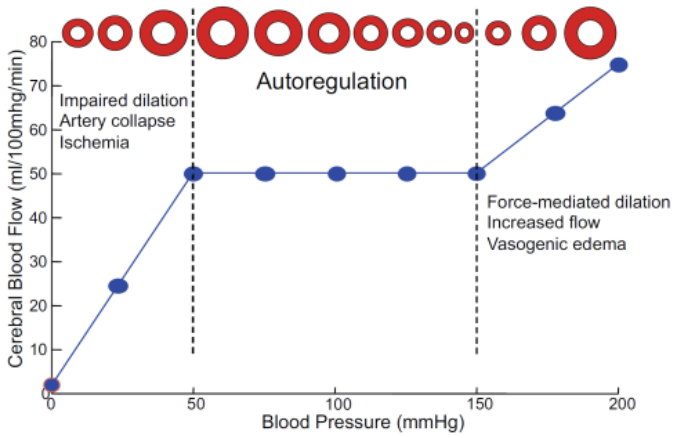


Cerebral metabolism:

Head injury results in an increase in the activity of excitatory neurotransmitters and cerebral metabolism which can result in neuronal death.

Autoregulation:

In a normal brain, cerebral blood flow is maintained over a wide range of blood pressures through cerebral autoregulation. However, autoregulation is lost in severe traumatic brain injury and as a result cerebral perfusion depends directly on the blood pressure which is why maintaining blood pressure in a target range becomes crucial (*Figure 3*).



Cerebral perfusion pressure (CPP):

CPP is calculated as the difference between MAP and ICP. A CPP of 40 mmHg in infants and 50 mm Hg in adolescents is considered optimal. CPP is optimised by lowering ICP and by maintaining an adequate MAP through vasopressors.

There are several interventions to lower ICP and are tiered based on ease of implementation, and effectiveness.

Tier I therapies include:-

1. Positioning :- Head end elevation to 30^0 to promote venous drainage. The head is kept in the midline to prevent unilateral jugular venous compression.

2. Sedation and intermittent paralysis

3. Normothermia /Prevention of fever

4. Maintaining target CPP; if ICP monitoring is not available, assume an ICP of 20- 25 mm Hg and target MAP accordingly.

5. Osmotherapy – 3% saline or mannitol

6. Neuroprotective ventilation- target pCO_2 35- 40 mmHg and paO_2 > 60 mm Hg with continuous pulse oximetry and end-tidal CO_2 monitoring.

Tier II therapies include:-

1. Barbiturates (phenobarbitone or thiopentone)

2. Decompressive craniectomy: unilateral or bilateral

3. CSF diversion (external ventricular drainage, or lumbar drainage)

Supportive measures

Maintenance fluids should be restricted to about 70- 80% of the calculated volume as these children invariably have SIADH and tend to develop fluid overload and hyponatremia. Isotonic fluids (normal saline +/- glucose or Plasmalyte) should be used as maintenance fluids. Early enteral nutrition should be started.

Case continues:

The child was ventilated for 48 hours after which sedation was stopped and neurology was assessed. As she was conscious and hemodynamically stable with good cough reflex and respiratory efforts, she was put through a spontaneous breathing trial and successfully extubated. Noradrenaline and 3% saline were stopped. She was discharged on levetiracetam. She child had transient truncal weakness for which physiotherapy was started. She was discharged after 7 days of hospitalisation. Truncal weakness had subsided at two-weeks' follow up.

Comments:-

Prevention of fever is extremely important as hyperthermia raises cerebral metabolism and increases excitotoxicity. However, therapeutic hypothermia is not routinely recommended. Recent guidelines suggest that moderate hypothermia may be used (Level III). Antiepileptics are recommended for the prevention of early (< 7 days) post-traumatic seizures but have no role in the prevention of late onset seizures. There is no role for the routine use of antibiotics or steroids in severe TBI.

Post ICU care plays a major role in the rehabilitation of such children. Occupational therapy, nutritional support etc are important for complete recovery.

Neurological impairment – motor, cognitive and speech- is often severe at hospital discharge. However, young children have a tremendous potential for a near complete recovery. Gradual recovery of neurological function- often over several months- is seen cases with adequate rehabilitation.

Conclusion:

Early and sustained stabilisation play a crucial role in improving survival and neurological outcomes in children with severe traumatic brain injury. Maintaining adequate cerebral perfusion, preventing further secondary injury and treating raised ICP are important aspects of the management during the acute ICU phase. With timely and appropriate therapy, children can make a good recovery.

References:

1. Kochanek et al. Guidelines for the Management of Pediatric Severe Traumatic Brain Injury, Third Edition: Update of the Brain Trauma Foundation Guidelines. Ped Crit Care Med 2019: 20: S1- S82

2. Furhman's and, Zimmerman's Pediatric Critical Care 5th Edition; Elsevier 2017.

3. Roger's Textbook of Pediatric Intensive Care 5th Edition; Lippincott Williams and Wilkins 2015.

A Doctor with a Mission

" the kind hearted, gentle and ever smiling Dr. Harivallabh P. Pai".

Born on the 13th of January 1958 at Hospicio Hospital Margao, Dr Pai was told he was a low birth weight baby and remained so till his college days. Sir did his schooling from St. Joseph's Institute Vasco, science degree from Chowgule College Margao, MBBS from Goa Medical College Bambolim and DCH (CPS) from Bombay.

RESIDENCY DAYS

After joining Paediatrics, during the first residential post, Dr. Pai got a call from KEM Hospital (Mumbai) head clerk informing him of his selection for the MS ENT post, which he had applied for. However, a few days into the field of Paediatrics and the world of children instilled a keen interest in him to work with children and he immediately dropped the idea of joining surgery. In the words of Dr Pai- "I found Paediatrics very challenging as children cannot express and we as Paediatricians need to solve the puzzle with the help of their parents."

Sir fondly remembers how lucky he was to be able to work under different HOD's in Mumbai and Goa including Dr Lokeshwar and Dr Laxmi Gaunekar who have helped him adopt 'The art of history taking' which, is the most important tool for a clinician.

HOBBIES

Acting : Sir won the 1st prize in pantomimic acting at All India Intermedical Youth Festival at Baroda. He has participated in many one-act plays during the college days.

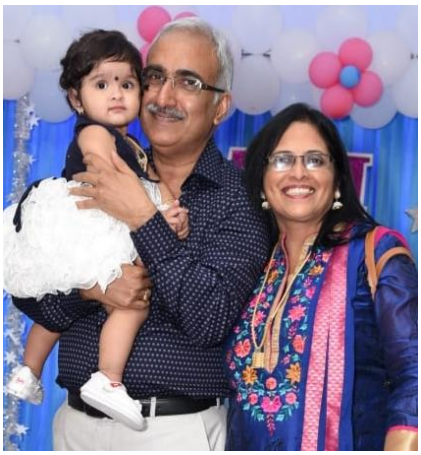
Sports : playing table tennis. Sir was runner up at the All Goa Intercollegiate TT tournament, held during MBBS days.

Music (listening and singing) is Sir's passion. He was one of the finalists at the "All Goa Senior Citizens" singing competition held this year.

Travelling with family, including attending all the national Pedicon conferences.

FAMILY

Dr. Pai is happily married to Dr Sunita who happens to be his classmate and an anaesthetist and a regular source of inspiration in his life. He is blessed with a son, Amey and his wife Sanjana. His daily source of energy is his 4year old granddaughter Saanvi who is 'the apple of his eye'.



PAEDIATRICS IN PRACTICE

Armed with the qualities of patience, compassion, humour and a lot of clinical knowledge and experience, Dr Pai set up his clinic in Vasco da Gama in 1986. He along with a group of doctors in partnership also started a multispeciality hospital "Sanjeevani" in 1995. After 20 years of successfully running the hospital, they leased it out to a third party due to retirement of the senior doctors.

In his professional career Dr Pai has been a humble but true leader.

He was - State IAP president (GAP) 2017-2018

- State IMA president (2012)
- State president of APNH (association of private nursing homes) Goa
- Founder member of GAP

He took initiative of organising Goa Pedicon 2018 as well as GIMACON in 2012.

His leadership qualities and enthusiasm date back to 1979 when he organized Gomefest in Goa. He was then the general secretary of the Gomefest (during 3rd MBBS), which is a "one of its kind" All India Intermedical youth festival.

During his tenure as IAP state president – Goa IAP won the best branch award consequently for 2 years. He was also felicitated by IMA Goa state in 2015 at Gimacon 2015 at the hands of the health minister.

Dr Pai has been in clinical practice for almost 34 years and although science has progressed by leaps and bounds, amidst the many gadgets, tests, scans and MRI, Sir feels that the very essence of a doctor – patient relationship ---'personal touch' is fading away. Sir is inspired by Dr Y.K Ambdekar(Ethical practice) and Late Manohar Parrikar (simple life with high level of thinking).

MEMORIES THAT HAVE CREATED AN IMPACT

Dr. Pai had adopted the practice of passing a feeding tube after every Caesarean section (taught to him by his HOD during PG days at Mumbai). Years ago, after struggling to pass a feeding tube in a baby whose delivery he had attended and finding a murmur on cardiac auscultation, Dr. Pai referred the baby to Criticare, Panaji, and after investigations the baby was diagnosed as a case of double TOF (Tetralogy of Fallot and Tracheo- oesophageal fistula). The baby received timely intervention and is today a healthy youngster.

On another occasion, Dr Pai was pleasantly surprised to " pop the champagne" at the 21st birthday of one of his patients.

The three most important virtues in life are "commitment, faith and benevolence." Dr Pai's advice to the upcoming young paediatricians is to be more empathetic, patient, always wear a smile, communicate what is important and take decisions ethically. At the same time he advises us to take a break, to not be a workaholic, take care of our health and spend quality time with family.

In the words of Dr. Pai "Never make false promises, but try your level best because वादे अक्सर टूट जाते हैं, कोशिशें कामयाब हो जाती हैं."

"Thalassemia" --- an update

-Dr.Lorraine D'Sa,

Lecturer, Dept. of Pediatrics, GMC.

Hemoglobinopathies, which include the thalassemias and structural hemoglobin (Hb) variants, are the most common group of autosomal recessively inherited monogenic disorders of Hb production and pose a significant health burden in India.

The prevalence of β-thalassemia carriers in the Indian population is 3-4%. Some ethnic groups like Sindhis, Kutchis, Lohanas, Punjabis, few Muslim groups as well as few tribal populations have a higher prevalence (5-17%).

Pathophysiology: The basic defect in β-thalassemia is a reduced or absent production of beta globin chains with relative excess of α-chains. This causes extensive premature destruction of the red cell precursors in the bone marrow and in the extramedullary sites referred to as "ineffective erythropoiesis" and is the hallmark of β-thalassemia.

Clinical presentation of β-thalassemia major usually occurs between 6-24 months of age depending on whether the individual genotype is β^0 / β^0 or β^+ / β^0 ; with severe microcytic anemia, mild jaundice and hepatosplenomegaly being the presenting features. Affected infants fail to thrive and become progressively pale.

β-thalassemia intermedia (β^+ / β^+) should be suspected in individuals who present at a later age with similar but milder clinical features.

β-thalassemia trait (β / β^+) has no clinical effects since the activity of the normal β gene makes enough stable hemoglobin.

Hematologic diagnosis of β-thalassemia major is characterized by reduced Hemoglobin level <7gm/dl, MCV between 50-70fl, and MCH between 12- 20pg. Peripheral smear examination shows microcytosis, hypochromia, anisocytosis, poikilocytosis, target cells and erythroblasts.

Qualitative and quantitative diagnosis of β-thalassemia major is made by identifying the amount and type of hemoglobin present in an individual by doing the Hemoglobin electrophoresis or high pressure liquid chromatography (HPLC) test. In β^0-thalassemia homozygotes, HbA is almost absent and HbF constitutes 92-95% of the total hemoglobin. In β^+ homozygotes or β^+ / β^0 genetic compounds, HbA levels are 10-30% and HbF between 70-90%. HbA2 is variable in β-thalassemia homozygotes.

However elevated HbA2 levels are diagnostic of β-thalassemia trait/ minor/ carriers.

Transfusion protocols

The recommended treatment for thalassemia major involves lifelong regular blood transfusions, to maintain the pre transfusion hemoglobin level between 9 – 10.5 g/dl. This transfusion regimen promotes normal growth, allows normal physical activities, adequately suppresses bone marrow activity in most patients and minimizes transfusional iron accumulation.

It is imperative however to confirm the diagnosis of beta thalassemia major before initiating a patient on lifelong transfusion therapy.

Following parameters suggest that the patient will need chronic red cell transfusions.

- Hb level <7 g/dl on two successive occasions separated by at least 2 weeks (the patient should be on folic acid replacement and there should be no other aggravating cause, i.e. infection, bleeding, etc.)
- Patient's growth, activity, academic performance, zeal, etc., are hampered
- Unnatural bony growth due to marrow expansion
- Development of organ failure such as cardiac failure, edema
- Even if Hb level is >7 g/dl and <10 g/dl, and above clinical features are present, the patient may need chronic transfusion therapy.

Objectives of chronic red cell transfusions

- To ensure adequate Hb level so that O2 delivery to the tissue is not hampered. This will be indicated by:
- Normal growth spurt
- Increased zeal, energy, enthusiasm, and improved academic purpose
- Improved appetite
- To suppress over active erythropoiesis leading to bone deformities.

Frequency of red cell transfusions

Red cell transfusions should be given at an interval of 2-5 weeks.

This interval is optimized based on:

- The amount of red cells transfused so that pretransfusion Hb remains >9 g/dl but post transfusion Hb does not go above 12 g/dl
- There is no fluid overload
- Transfusion process is over within a reasonable time (4-6 h)
- Frequency of transfusions is not such that it interferes with patient's normal activities
- Reducing the number of Venipunctures (as lifelong transfusion is needed, peripheral veins need to be preserved well).

How much red cell to be infused?

- Pretransfusion Hb is to be estimated
- Weight of the patient is to be recorded
- If the HCT of the red cell concentrate used is 0.65, then 3-4 ml/kg will raise the Hb by 1 g/dl in the absence of hypersplenism
- Generally in a single transfusion an attempt is made to raise the Hb by 4 g/dl if transfusions are scheduled at 3- to 5 weekly intervals.

Suitable blood products for transfusion

Careful donor selection and screening, favouring voluntary, regular and non-remunerated blood donors is ideal. Before the first transfusion, extended red cell antigen typing of patient at least for C, E and Kell is required. Transfusion of ABO, Rh(D) compatible blood matching for C,E and Kell is recommended. Red cell concentrates with hematocrit around 0.65 are suitable. Leukoreduced packed cells are ideal to reduce incidence of transfusion reactions. Saline washed red cells are required for patients who have severe allergic reactions.

Blood transfusion exposes the patient to a variety of risks and adverse events including non-hemolytic febrile transfusion reactions, allergic reactions, acute hemolytic reactions, alloimmunisation, delayed transfusion reactions, autoimmune hemolytic anemia, transfusion related acute lung injury and transfusion induced graft versus host disease.

Management of iron overload

The cause of iron overload in thalassemia patients is both due to the regular red blood cell transfusions and increased absorption of iron through the GI tract. Chelation therapy aims to balance the rate of iron accumulation from blood transfusion by increasing iron excretion in urine and/or faeces with chelators. The methods used to quantify the amount of iron overload include serial measurements of Serum ferritin levels, liver iron concentration measurement by liver biopsy, magnetic biosusceptometry (SQUID) or by T2* MRI. Estimating serum ferritin levels is readily available and easy to monitor. When the level is above 1000 ng/ml, chelation therapy should be initiated. Two oral iron chelators are available in the market at present, Deferiprone (DFP) and Deferasirox (DFX). At present, both iron chelators are found to be effective. While DFP increases excretion of chelated iron through urine, DFX increases excretion through both stool as well as urine.

DFP is given at a dose of 75 mg/kg/day in three divided doses. DFX is given at 30 mg/kg/day in a single dose after dissolving the tablet in water or orange/apple juice. It has been observed that Indian children require Deferiprone in a dose of 100 mg/kg/day while Deferasirox in a dose closer to 40 mg/kg/day to lower serum ferritin levels.

The action of iron chelators is best on an empty stomach.

The injectable iron chelator is desferrioxamine and it has been used for the longest duration. It is given as a subcutaneous injection over 8-14 hrs, 5 days a week at a dosage of 40 mg/kg/day. Combination regimen using DFO and DFP or DFX is usually recommended for patients with high serum ferritin levels and patients having cardiac and liver iron overload and in children with endocrine problems.

Monitoring

Regular monitoring of thalassemic children is of utmost importance in daycare centers with the following objectives:

- To ensure that pre transfusion Hb is maintained between 9 and 10 g/dl
- To detect development of leucopenia or thrombocytopenia
- To monitor iron status by serum ferritin and MRI T2*
- To detect the side effects of transfusion or complications of chelation therapy early.
- Early detection and management of transfusion transmitted infections
- Monitoring of cardiac functions by X-ray chest, Echo, or MRIT2* images if required.
- To monitor growth and development and institution of therapy at the earliest for growth failure
- Early detection of endocrine problems and institution of appropriate management.

It is desirable that all the clinical and laboratory parameters of the patient should be recorded on a predesigned proforma. Anthropometric measurement should be properly recorded using the growth charts.

Regular monitoring of thalassemic children is helpful in preventing both complications of the disease as well as of the therapy. It helps in early detection of growth failure, development of endocrinopathies, liver, and cardiac complications. Appropriate and prompt management can reverse most of the complications.

Bone marrow transplantation

It offers a permanent cure and better future for children. The credit of the first bone marrow transplantation (BMT) in thalassemia major goes to E Donald Thomas who performed this procedure in an 18-month-old thalassemic child in 1982 using a HLA-matched elder sister as the donor. This child was cured of thalassemia!

The cost of BMT in India is around 8-10 lakhs and is regularly being done at Christian Medical College Vellore, Tata Memorial Hospital Mumbai, All India Institute of Medical Science (AIIMS) in New Delhi, Sanjay Gandhi Postgraduate Institute of Medical Science (SGPGI) Lucknow and Post Graduate Institute of Medical Education and Research (PGI) Chandigarh. Several other centers' in corporate hospitals are providing bone marrow transplant facilities.

All thalassemic children should be considered for BMT if a HLA-matched donor is available. Several non-government organizations (NGOs) and state governments are providing financial assistance. Cost of BMT is a onetime expense and its results are far better than conventional therapy.

Thalassemia management in Goa

The department of Pediatrics at Goa Medical College Bambolim, manages around 80 patients with transfusion dependant anemias.

The hypertransfusion protocol is followed and all patients are encouraged to maintain a pre transfusion Hb of 9 -10.5 gm%.

ABO and Rh matched packed RBC transfusions through bedside WBC filters are provided @ 15-20ml/kg every 2-4 weekly for all patients.

Chelating agents i.e. Desferrioxamine, Deferiprone and Deferasirox are also provided along with Folic acid, Calcium and vitamin D supplements. Iron overload is monitored by serial Se.Ferritin assessments every 3-4 monthly. Growth monitoring and monitoring of blood counts, RFT's and LFT's is done 3-6 monthly. 2D ECHO, Abdominal USG, Thyroid function tests, HIV, HbsAg and HCV tests are done yearly for all patients. Endocrine evaluation is also done for all adolescent patients.

Thalassemia society of Goa

The "Thalassemia society of Goa", (Reg No.-388/GOA/2015) registered in October 2015, is a purely voluntary, social welfare society, committed for the cause of Thalassemia and endeavors to support the treatment, care and cure of poor and needy thalassemics.

The society's mission is :

1. To ensure free and safe availability of blood, leucocyte filters, chelation drugs and investigations for all Thalassemic children.

2. To conduct carrier detection camps to ensure zero births of Thalassemic children by providing counselling of carriers prior to their marriage.

3. To provide genetic counselling to couples who are already carriers.

4. To provide counseling to children affected with Thalassemia to help them cope with the disease.

The society's vision is :

1) To see children with Thalassemia lead as normal life as possible.
2) That through awareness we see the incidence of this disease decline considerably.

Over the last 3 years, the society has organized CME's for doctors inviting noted hematologists like Dr.Mamta Manglani and Dr.Rashid Merchant to examine and opine regarding care of thalassemics. The society also supported DEXA scans and T2* MRI for older patients. The society organized an educational tour cum get-together for the children and parents, to encourage bonding and fellowship amongst the families.

Awareness campaigns through participation in the Rotary Rain Run, conducting awareness talks in schools, colleges, corporate offices, etc. were conducted. The most important achievement of the society being the HLA typing camp for thalassemia children below 14 years of age with healthy siblings to look for HLA matched siblings who could be the potential bone marrow donors for their affected sibling. Through this camp it was possible to know that there are 11 children who have HLA matched siblings. Most of these parents are willing to go ahead for the BMT of their child and the society is currently involved in trying to arrange funds for the same.

The way forward…

The incidence of Thalassemias can definitely be reduced by increased awareness of this condition in society. Due to migration and population admixture, Hb variants and the compound heterozygous conditions (Hb Sβ thalassemia, Hb E–β thalassemia, Hb D–β thalassemia) are now seen all over the country. Hence it is not sufficient to target any particular region.

Individual groups where screening and diagnosis could be done include: Screening of the extended family members of children diagnosed with thalassemia major, Pre-marriage screening in colleges/corporate companies, Antenatal screening and Preconception screening if both partners are carriers.

A joint effort by the State Government supported by the Centre along with help from NGOs, Thalassemia societies and corporate houses as a part of their social responsibilities and strongly backed by political will, would be required for successful implementation of a national control programme. Guidelines for this have recently been prepared by the National Health Mission, Ministry of Health and Family Welfare with the help of several experts in the country; and are awaiting implementation at the earliest.

References:

1. Kanjaksha Ghosh, Roshan Colah, Mamta Manglani, et al. Guidelines for screening, diagnosis and management of hemoglobinopathies. Indian J Hum Genet. 2014 Apr-Jun; 20(2): 101–119.

2. Cappellini MD, Cohen A, Porter J, Taher A, Viprakasit V . Thalassemia International Federation. Guidelines for management of Transfusion Dependant Thalassemias. 3rd Edition.

3. National Health Mission Guidelines on Hemoglobinopathies in India Prevention and control of hemoglobinopathies in India-Thalassemia, Sickle cell disease and other variant hemoglobins. Ministry of Health and Family Welfare, Govt of India; 2016.

Thyroid hormone transport proteins

-Dr Ramu Shashtri

MD paediatrics, PDCC (paediatric endocrinology)

Understanding thyroid hormone transport proteins and their role in interpretation of thyroid function tests is very important in clinical practice.

T4 and T3 circulate bound to transport proteins - TBG, albumin and transthyretin. TBG has high affinity and low capacity for binding while albumin has high capacity and low affinity. These proteins not only help in transport of the hormone but also act as a reservoir to stabilize the hormone levels. Only 0.3% of T4 and 0.02% of T3 circulate in their free form. T4 is predominantly bound to TBG, while T3 binds to all three proteins. The binding of T3 to TBG is decreased in hypothyroidism increasing free T3 levels providing a defence mechanism against the adverse effects of low T3. TBG in increased in hyperestrogenic states (pregnancy, oral contraceptives and SERM use) causing increased total hormone levels. Free thyroid hormone levels are normal in this setting.

Thyroid hormone requirement increases with OCPs and pregnancy due to increased binding of exogenous thyroid hormones to the binding globulins.

TBG deficiency is an antosomal recessive condition associated with reduced total but normal free thyroid hormone. It should be suspected in clinically euthyroid children with low total T4 levels.

Spuriously low T3 and T4 levels with normal free thyroid hormones are observed in hypoalbuminemia (Chronic liver disease or nephrotic syndrome) or decreased TBG levels (testosterene and glucocorticord use).

Drugs inhibiting binding of T4 to TBPs (Heparin, NSAIDS, anti- Epileptic Drugs) can present with transient increase in FT4 levels with normal TSH levels.

Thyroid hormone transport proteins:-

a) Increased – High T4, Normal FT4, TSH

-- estrogen, OCs, Pregnancy, SERM

--Neonates

--Hepatitis, Porphyria

--Mitotane

b) Decreased – Low T4, Normal FT4, TSH

- low albumin—Nephrotic Syndrome , Chronic liver disease

-Androgens, Cortisol, anabolic steroid

-Nicotinic acid, L- asparginase

-Severe illness

c) Binding inhibitor- Normal T4, High FT4

-Salicylates

-Furosemide

- NSAID

- Phenytoin, CBZ

NEUROBLASTOMA WITH INCIDENTALLY DISCOVERED CONGENITAL HEART DISEASE

Dr Neha Da Rocha, Dr Anagha Dubhashi, Dr M.P. Silveira, Dr. Nikhila Gaude, Dr.R.G. W Pinto, Dr Suresh Mandrekar, Dr Sita

CASE REPORT

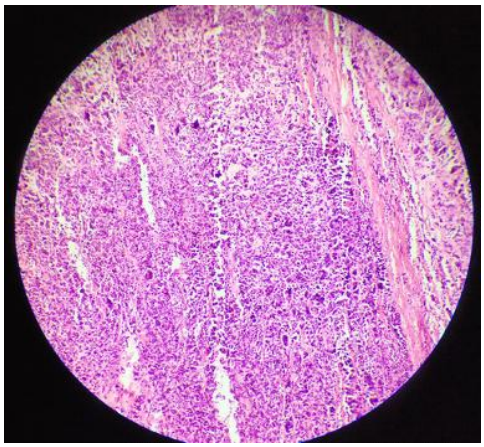
2 year old, developmentally normal female child born out of 3rd degree consanguinous marriage from UP, with no relevant family history and apparently alright till 15 days prior, came with chief complaints of abdominal pain and abdominal distension for 15days.
The abdominal pain was periumbilical in location, severe in intensity and intermittent.
The abdominal distension was generalised, gradual and progressive.
There is also history of constipation and irritability for the last 15 days.
There is no significant past history.

On examination

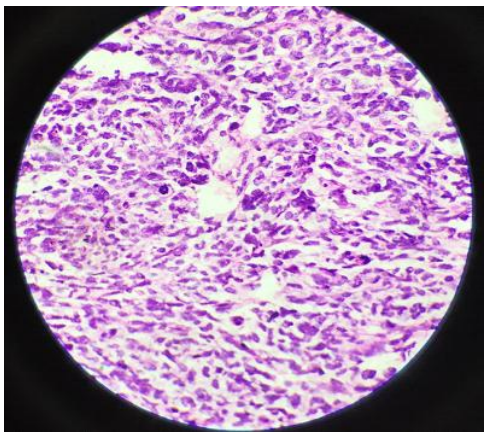
- Heart rate and Blood pressure were within normal range. Left pre-auricular lymph-node was palpable.
- Abdomen was uniformly distended. No liver or splenic enlargement. A hard mass was palpable in the left hypochondrium and lumbar region with irregular surface and margins. Systolic murmur was heard on auscultation.
- Blood investigations showed evidence of iron deficiency anemia, raised serum LDH (>3325) and elevated serum uric acid (6.2). Urinary VMA levels were elevated.
- Plain CT Abdomen and Chest revealed a heterogeneous mass measuring 8*8 cms in relation to upper pole of left kidney, displacing the kidney caudally with specs of calcification suggestive of neuroblastoma .Extensive retroperitoneal adenopathy was present. Loculated collection possibly blood, in epigastrium between nodes and left lobe of the liver was seen.
- The child's sensorium worsened and was shifted to PICU and ventilated. Inotropes were added in view of progressive shock. Transfused two packed cells. 2D ECHO showed dilatation of all 4 chambers of the heart with LVEF of 20%. On 6th day of admission child went into multiorgan dysfunction and expired.
- Post Mortem examination showed Anaplastic neuroblastoma of left adrenal involving retroperitoneal lymph nodes infiltrating into the left kidney and forming metastatic nodules in the left kidney and pancreas along with bone marrow metastasis. Incidentally aortic stenosis with post stenotic dilatation

was discovered (probably congenital) along with chronic passive venous congestion in lungs and liver. This gets classified as a stage 3 neuroblastoma.

Left kidney with adrenal mass



Sheets of round tumor cells 10*



Tumor cells showing scanty cytoplasm and vesicular nuclei 40*

Incidentally discovered aortic stenosis with post-stenotic dilatation

DISCUSSION

Neuroblastoma is an embryonal cancer of the postganglionic sympathetic nervous system, which mostly arises in the adrenal gland. It is the most common extracranial solid tumour in children, comprising 8% to 10% of all childhood cancers. The incidence is nearly one per 10,000 children under the age of 15 years.[1]

The association between congenital heart disease and neuroblastoma has been described, with different theories put forward to explain this relation.[2]

This association is considered to be plausible as neuroblastoma originates from embryonal neural crest-derived cells and neural crest-derived cells are essential in cardiogenesis as well. Neural crest cells play an important role in the septation of the outflow tract of the heart and in the formation of the conotruncal part of the ventricular septum.[1]

Neuroblastomas: dominant associated cardiac malformation	Total	In situ	
		No	Yes
Persistent arterial duct	9	5	4
Transposition of the great arteries	8	4	3
Ventricular septal defect	8	3	3
Hypoplastic left heart syndrome/mitral atresia	5	2	3
Tetralogy of Fallot/pulmonary atresia	6	3	2
Aortic stenosis/bicuspid aortic valve	4	1	2
Coarctation of aorta	4	3	1
Anomaly of aortic arch/vascular ring	3	1	2
Pulmonary valvar stenosis	2	2	0
Cor triatriatum	2	0	0
Atrial septal defect	2	0	1
Atrioventricular septal defect	2	1	0
Complex cardiac malformations	5	2	3
Unclassified and others	7	6	1
Total	67	33	25
Total neural crest derived anomalies	21	10	9

Table shows the reported cases of neuroblastoma and congenital cardiac disease as published in Archives of Diseases in Childhood in 2002.[2]

George and colleagues demonstrated that children affected with NB have a higher prevalence of CHD; however, van Engelen and colleagues have denied evidence of association between these two conditions. A review of more than 1900 cases showed that NBs account for approximately 17% of the malignancies seen in Costello and Noonan syndromes, a disorder characterized by diverse tissue and organ defects, including CHD.[3]

However,the association between neuroblastoma and CHD still remains unclear. To confirm or reject the true existence of such an association, further research in a large and complete cohort of neuroblastoma patients is needed. Standard cardiac screening in all patients with neuroblastoma is therefore not supported by current evidence.[1]

References.

1. Klaartje van Engelen et al. Prevalence of congenital heart defects in neuroblastoma patients: a cohort study and systematic review of literature. Eur J Pediatr. 2009 Sep; 168(9): 1081–1090. doi: 10.1007/s00431-008-0891-y
2. R Holzer,RCG Franklin. Congenital heart disease and neuroblastoma: just coincidence? Arch Dis Child 2002;87:61–64
3. Alessandro Testori et al.Exploring Shared Susceptibility between Two Neural Crest Cells Originating Conditions: Neuroblastoma and Congenital Heart Disease. Genes 2019, 10, 663. doi:10.3390/genes10090663

Breastfeeding..... an emotional attachment that gifts a lifetime.

- **Margaret D'Mello**
- **Sister in charge, SNCU, Hospicio Hospital , Margao**
- **Member of breastfeeding network of India**

Breastmilk is the best and complete food for the baby's nutrition in the 1st 6 months of life. It is currently accepted as the most important preventive step in promoting good health and protecting against disease.

As we all know breastmilk is an immune booster and the 1st vaccine to a baby. There is much evidence on the great potential of " these drops of milk" in protecting and promoting early recovery from serious illnesses. But, what is often forgotten or not emphasised upon are the long term benefits of breastfeeding, which is why it is now recognised as " the need of the hour" . It is important to see how exclusive breastfeeding for the 1st 6 months provides a shield against diabetes, hypertension, heart disease, asthma and allergies. Although these chronic diseases manifest at a later age, they have their onset right from infancy. Breastmilk also provides a protective environment in which the brain grows and is the best prevention against mental illnesses and poor emotional development. So.... in all Breastfeeding is the foundation of good health.

Breastfeeding appears so natural, yet millions of babies lose the battle because of failure to breastfeed.

What are the impediments to successful breastfeeding ?

There are many causes but all with solutions

Lack of knowledge, myths/cultural practices, "Not enough milk" phenomenon, work place related constraints.

The most important factor to successful exclusive breastfeeding is the parents–baby relationship and hence the desire/commitment to give the baby their best. I am happy that this year the WHO theme for the world breastfeeding week is "EMPOWER parents, ENABLE breastfeeding now and for the future".

It is the collective effort of the new mother and her family, remembering that the medical fraternity is always there to help them master the art and to answer their queries and alleviate their fears.

How can we as medical professionals help?

-Checking mothers antenatally and those that are likely to have difficult latching for e.g inverted nipples have to be counselled and marked with a star so that at delivery these babies and mothers are attended to.

- Providing correct and complete information regarding advantages and technique, including trouble shooting.

- Giving babies born by Caesarean section their first breastfeed in the operation theatre.

- Encouraging Rooming–in and Kangaroo mother care in low birth weight babies.

- Be prepared to answer any questions that mother's have which will increase their confidence in breastfeeding.

So let us set our priorities right, sensitise families and parents about this beautiful gift, which is the best investment and health insurance for a child.

In all my years of service, I have seen the struggles, comforted the tears, felt their pain but through it all ,with support and encouragement I have seen a joyful smile once again, not only on the face of the mother but also the child who grows up to be a healthy infant.

Breastfeeding is like a new job....

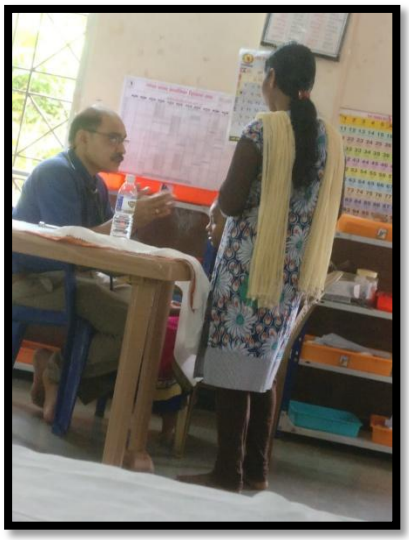
Its' hard in the beginning....

It's easier with practice....

And you don't dare quit....

Because you will miss out on all the rewards.

ACTIVITIES OF JULY TO SEPTEMBER



Medical camp was organised in July 2019 at Kukkali along with Balaji and Sairam trust. Dr Anant Kini, Dr Rajendra Dev and Dr Sharad Raikar participated in the camp. 90 Children were seen and the common problems identified were undernutrition, anemia ,dental caries, wax in ears, visual defects, scabies. Dr Sharad spoke to parents about importance of oral electrolyte solution, how to make home based ORS and cleanliness. Miss Nikisha Shetye was present who advised about diet. Dr Vaishali Kerkar, dental surgeon was a guest of our team who advised children about dental care.

Dr Sumant Prabhudesai attended the TOT programme on IAP ALS Mass Awareness held at Pune on 14th July 2019

CIAP Programme raising awareness on newborn screening held at Goa Medical College ENT auditorium on 28/07/2019. Speakers were Dr. Ambiprasad Mohite and Dr. Kumar S.R.

Topics covered:

a) Need for early detection of hearing impairment.

b) Details on OAE test and how it works

c) Demonstration

Dr Poonam Sambhaji took part in the breastfeeding week celebration at DIET(District Institute of Education and Training) College Porvorim on the 7th of August. The audience consisted of 120 students in the age group of 18-28 years.

Dr Poonam Sambhaji participated in the breast feeding week celebration at RG stone hospital Porvorim on 6th August 2019

BREASTFEEDING WEEK 1ST TO 7TH AUGUST 2019.

Asilo hospital

https://www.dropbox.com/s/5r2o3u3y8m3a48e/bfw%20%20prog.%20video.mp4?dl=0

The world breastfeeding week was celebrated at North District (Asilo) Hospital Mapusa from 1st to 7th August 2019. The following is the report of the activities carried out.

Poster competition:

A ward wise poster competition was held on the 1st of August. Participants were all staff nurses. A total of 16 posters entered the competition.

The competition was judged by Dr. Shweta Gandhi and the Assistant Matron.

The posters were awarded marks for correctness of information, representation of this year's theme and creativity.

These posters were on display through out the breast feeding week in the hospital lobby.

Paylo ghans / first food competition:

Ward wise cooking competition was held on 1st august.

The participants were all staff nurses. They were to prepare complementary foods suitable for infants 6-8 months of age.

The dishes were judged by Dr. Chetna Khemani and Ward in charge SNCU Sister Milagrosa Rebeiro.

The dishes were awarded marks for consistency, nutrient value and palatability and use of locally available ingredients.

What's in my tiffin competition:

Another cooking competition was held on 2nd august.

The participants were all patient attendants. They were asked to cook dishes which children would love in their tiffins.

The dishes were judged on nutrient value, taste, presentation and ease of preparation (as tiffins are always made in a hurry).

The dishes were so good we were forced to give out 5 prizes in place of 3.

Talk in ANC OPD and pediatric OPD:

A talk to sensitise pregnant women on the need for and the importance of breastfeeding emphasizing on the supportive role of the father was held in the waiting area of the ANC OPD and paediatric OPD.

Our breast feeding counsellor Mrs. Mithila Kamat gave the talk.

Breast feeding support group:

As they say it takes a tribe to raise a child. In today's scenario joint family and robust support systems for breastfeeding and child care are lacking. New mothers therefore feel lonely and have a lot to share.

So we choose this activity were mothers in our postnatal ward were divided into groups were they could share their queries, concerns and experiences with each other.

This group discussion was facilitated by our SBA students and supervised by Mrs. Mithila Kamat our breast feeding counsellor.

Skit on breast feeding by Institute of Nursing Education students:

Students of INE Bambolim performed a skit on Breast feeding in the paediatric OPD and the post natal ward.

Skit by Eco- clean staff:

The Eco- Clean staff of the hospital put up a skit on breastfeeding in the post natal ward.

Skit by Vrundavan school of nursing:

A skit on breastfeeding was performed by Vrundavan Institute of Nursing Education in the antenatal OPD and the post natal ward.

It was very innovative. They even performed a puppet show.

Passing the parcel game:

A passing the parcel game was held in the post natal ward for all the mothers. The forfeit was to answer questions on breastfeeding. Three prizes were awarded.

Sensitization on breastfeeding in private nursing homes:

Our staff nurses sister Clevy D'souza and Sister Arpita Mayekar went to various private nursing homes and gave talks to sensitize mothers who delivered in private hospitals about the need for and the importance of breastfeeding.

Talk on breastfeeding at Doordarshan:

Our Dr. Preeti had a live phone in programme on Goa Doordarshan to spread awareness on breastfeeding and infant and young child feeding.

Distribution of galactogoges:

The Inner Wheel Wing of the Rotary Club of Mapusa distributed galactogouges to our post natal mothers.

Talk on breast feeding:

A talk on breast feeding was given by Dr. Swechha Kamat. This talk was very informative as she showed a video of "breast crawl".

Valedictory function and Prize distribution Ceremony:

A valedictory function was held on the 7th of August.

Dr.Lalita was the chief guest and our MS Dr. Mohandas Pednekar was the guest of honour.

Prize distribution for all the competitions that were held during the week was held as part of this function.

The winners of the cooking competitions presented their dishes to our esteemed guests.

HOSPICIO HOSPITAL CELEBRATES

WORLD BREAST FEEDING WEEK AUGUST 1ST TO 7TH 2019

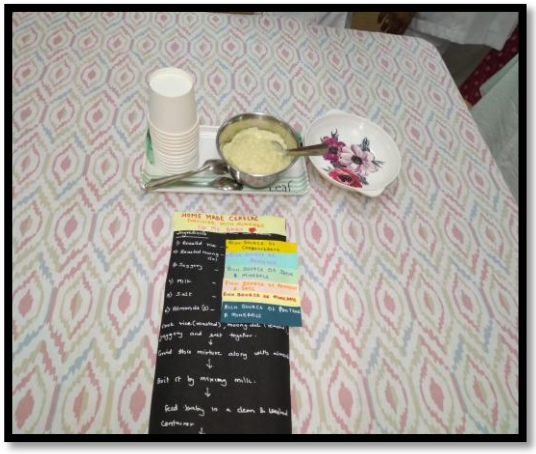
Please click on following link for a detailed presentation

https://www.dropbox.com/s/y51f3inawu19nl4/breast%20feeding%20week.pptx?dl=0

Subdistrict Hospital Ponda celebrates breastfeeding week 2019

Breastfeeding Corner

Competition on weaning foods




RICE PORRIDGE
INGREDIENTS
NUTRITIONAL VALUE OF
RICE PORRIDGE
1 Bowl = 327 calories
Total Fat = 6.2 gram
Protein = 14.5 gram
Carbohydrates = 54.1 gram
Vitamin A 214 µg
Vitamin B6 0.2 mg
Vitamin B12 1.7 µg
Vitamin C 0.5 mg
Vitamin D 44 µg


HEALTHY WEANING FOOD FOR BABIES
FROM: 6 MONTH - 2 YEARS
BEST FOODS FOR BABY LED WEANING
DRY FRUITS POWDER
VEGETABLE SOUP
VEGETABLE SOUP
RICE & MOONG DAL POWDER
APPLE PUREE
HOMEMADE PORRIDGE
HOMEMADE CERELAC
RAGI PORRIDGE


BEGIN TO FEED AT 6 MONTHS
FROM 7-8
FROM 9-12
FROM 13-14
MIXED PUREE
INGREDIENTS:-
WASH YOUR HANDS
USE CLEAN UTEN


WASH YOUR HANDS BEFORE COOKING
USE CLEAN UTENSILS BEFORE COOKING
HEALTHY WEANING FOODS FOR BABIES

Exhibition on promotion of breast feeding

Hand painting competition on promotion of breast feeding

Talk by MO to postnatal patients on breast feeding

Goan representation at 'Neuropedicon 2019' at Hyderabad

Dr Vibha Parsekar presented a poster based on evaluation of parent training programme for parents of children newly diagnosed with autism.

Dr. Ashwin Sardessai awarded the 2nd prize for platform paper presentation at Neuropedicon 2019, National Conference of IAP Paediatric Neurology Chapter. The paper presentation titled- Clinical profile of children with Congenital Muscular Dystrophy at a tertiary care referral centre in Southern India.

He also presented a poster of a case series of paediatric patients with Van der Knapp disease at the same conference.

Dr Sushma Kirtani , as faculty at **National Conference of IAP Adolescent Chapter 2019.**

Topic of discussion: Menstrual issues in adolescent girls.

Dr Sushma is also nominated by adolescent health academy on the editorial board of Indian journal of adolescent medicine.

Felicitation of Dr. Avadhut Kossambe at the hands of Dr Claude Billeaud, President de I'AEEP, France and Dr IndraShekhar Rao, Navodaya Hospital Hyderabad at World Paediatric Conference in Singapore on 10th September 2019.

Live phone in programme on doordarshan on deworming by Dr. Swechha Kamat

Live phone in programme on Rotaviral launch in the immunisation schedule in Goa by Dr. Anuradha Ghanekar.

Dr. Anuradha also participated in the Media Sensitisation Workshop on introduction of Rotaviral vaccine along with other members of the Directorate of Health Services.

5th Paediatric Mechanical Ventilation Workshop held at Goa Medical College

The Department of Paediatrics Goa Medical College along with the IAP college of Paediatric Critical Care organised this workshop on the 24th and 25th of August 2019 under the leadership of Dr Mimi Silveira . Dr Soonu Udani, Dr Madhumati Otiv, Dr Mahesh Mohite, Dr Sumant Prabhudesai, Dr Abhishek Chavan and Dr Iragouda Patil were the faculty. The workshop was attended by around 32 delegates. There were lectures followed by workstations where the delegates got hands on training regarding the various aspects of invasive and non-invasive ventilation.

Paediatric Neuro- rehab centre organised a CME on Paediatric audiology

The CME was held on the 19th of July in th GMC library auditorium
The speaker Dr Vikrant Patil, Dr of audiology spoke about hearing loss and intervention. Dr Archan Naik , consultant in ENT and cochlear implant surgeon spoke about cochlear implant surgery in hearing loss patients.

STEER Programme held on 15th September 2019

Faculty for the Programme were Dr Y K Amdekar , Dr R R Chokhani and Dr Harshad Kamat.

The Topics covered included

-Rational approach to history, examination and diagnosis

-When to act/ observe / investigate/treat

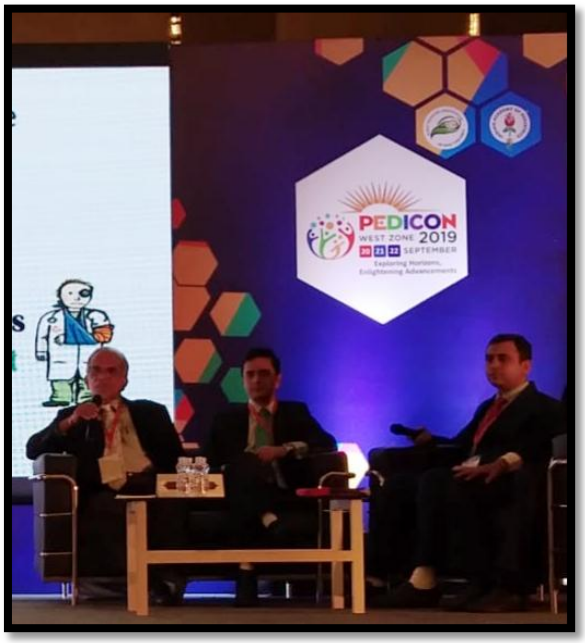
- How to make a provisional diagnosis

- Attempt anatomical/pathological/ etiological/functional diagnosis

- Common cases of fever and cough- seen in routine office practice

- Common cases of diarrhoea, jaundice, anaemia , renal problems

Dr Avadhut Kossambe as panelist for discussion on 'Medico-legal issues in Paediatrics' at **west zone Pedicon Surat** on 21st September 2019



BLS/ALS held in GMC on 21st and 22nd Sept 2019

The IAP BLA/ALS course was organised by department of paediatric GMC in association with IAP Goa state chapter. 36 delegates including paediatric residents and private paediatricians registered for the same. The course director was Dr Manish Doshi from Mumbai long with other instructors Dr Anjali Oti, Dr Geeta Bhat, Dr Hrishikesh Dingankar, Dr Nivedeta Patil and Dr Jaisinh Raorane. Dr Lorraine D'sa was the course coordinator. All the delegates found the course very helpful and useful in their clinical practice.

Dr Chetna Khemani as a panelist on ‘Mimics in rheumatology’ at National **conference of paediatric rheumatology** held on 28th and 29th September in Chennai

Dr Poonam Sambhaji took part in a camp conducted by rotary club Porvorim where she examined 30 children on 29th September 2019.

1. All of the following are true statements about Vi polysaccharide typhoid vaccine except
   a) TCV typhoid is preferred over Vi Polysaccharide vaccine
   b) T cell independent
   c) Interferes with Widal test
   d) Needs to be repeated every 3 years.

2. Chlorine added to swimming pool water cannot kill which of the following organisms
   a) E.coli
   b) Enterovirus
   c) Cryptosporidium
   d) None of the above

3. Short 4th metacarpal is seen in the following condition
   a) Congenital hypothyroidism
   b) Pseudo hypoparathyroidism
   c) Down's syndrome
   d) Edward's syndrome

4. A 6 year old with Hb A-30%, Hb S-60%, Hb F-15%, Hb A2-5%, Hb10gm/dl and MCV of 60%.
   Which of the following is the correct interpretation
   a) Homozygous sickle cell anemia after transfusion of RBCs
   b) Homozygous sickle cell anemia on hydroxyurea
   c) Sickle β thalessemia
   d) Sickle trait

5. In case of intraocular posterior segment cysticercosis, what should be the immediate treatment of choice?
   a) Praziquantel
   b) Albendazole with steroid
   c) Oral steroid
   d) Surgical excision of cysts

6. In a well looking newborn male with hyperpigmented scrotum, which investigation will you do urgently?
   a) Electrolytes
   b) 17 OHP
   c) Blood sugar
   d) None

7. In a child with sudden infant deaths and cardiac rhythm abnormalities in some family members, which drug would you best avoid to give for cough
   a) Phenylephrine
   b) Pseudoephedrine
   c) Dextromethorphan
   d) Chlorpheneramine maleate

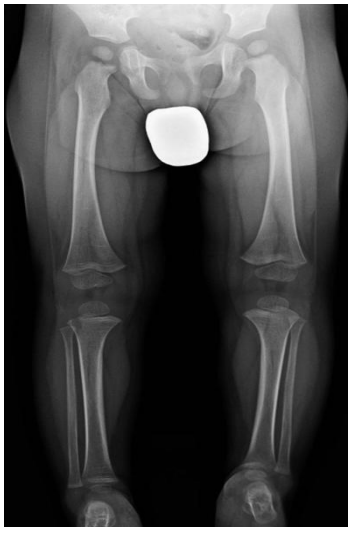
8. 

This is a 7 year old girl who presented to the paediatric OPD because of behavioural problems- being inattentive, and hyperactive with mild mental retardation.

Identify the syndrome:_____

9. **An 18-month-old boy:**

- With h/o limping for 1 week
- One episode with fever 10 days ago
- No articular swelling is observed



Differential diagnosis includes:

(Multiple answers might be correct.)

a) Osteomyelitis
b) Acute arthritis
c) Eosinophilic granuloma
d) Blount disease (tibia vara)
e) Fibrous dysplasia

10. Watch the clinical video
https://www.dropbox.com/s/44n99h3w5tl6nit/quiz%20video.mp4?dl=0

What is the diagnosis? Select ONE

a) Angelman syndrome
b) Cerebellar ataxia
c) Autism
d) William's syndrome
e) Noonan's syndrome
f) 22q 11 deletion syndrome
g) Rett syndrome

Kindly mail your answers of the quiz to dr.celineandrade@gmail.com. Special prizes await our first 3 correct entries.

Winners of July 2019 e-bulletin Quiz: Dr Annely D'Lima, Dr Sumant Prabhudesai and Dr Suvarna Naik.