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Editorial

Whole Fruit Versus Fruit Juice

Children can easily drink a lot of juice because the tastes of juices are good and Juices are easily carried around during the day as they are conveniently packaged or can be placed in a bottle or transportable covered cup.

Too much consumption of juice in a child's diet can contribute to different problems, like poor nutrition, obesity, and tooth decay.

Juice and vitamin

Juice contains a small amount of protein and minerals. Juices fortified with calcium have approximately the same calcium content as milk but lack other nutrients present in milk. Some juices have high contents of potassium, vitamin A, and vitamin C. In addition, some juices are fortified with vitamin C. The vitamin C and flavonoids in juice may have beneficial long-term health effects, such as decreasing the risk of cancer and heart disease.

Drinks that contain ascorbic acid consumed simultaneously with food can increase iron absorption by twofold. This may be important for children who consume diets with low iron bioavailability

Grapefruit and drug

Grapefruit juice contains substances that suppress a cytochrome P-450 enzyme in the small bowel wall. These results in altered absorption of some drugs, such as cisapride, calcium antagonists, and cyclosporin. Grapefruit juice should not be consumed when these drugs are used.

Fruit juice and diarrhoea

Toddlers' diarrhoea is a well-known and benign condition that often responds by simply removing excess juice from the diet of 1- to 4-yearold child..

However, malabsorption of carbohydrate in juice, especially when consumed in excessive amounts, can result in chronic diarrhoea, flatulence, bloating, and abdominal pain.

Fructose and sorbitol have been implicated most commonly but the ratios of specific carbohydrates may also be important. The malabsorption of Carbohydrate that can

result from large intakes of juice is the basis for some health care providers to recommend juice for the treatment of constipation, particularly in infants.

The North American Society of Paediatric Gastroenterology,Hepatology, and Nutrition constipation guideline suggests taking advantage of the sorbitol and other carbohydrates contained in some juices, such as prune, pear, and apple juices, to help increase the frequency and water content of stools for infants with constipation.

Unpasteurized juice products may contain harmful bacteria

Parents need to be informed that unpasteurized juice products may contain pathogens, such as Escherichia coli, Salmonella species, and Cryptosporidium species, which may be harmful to children. These organisms are associated with serious diseases, such as haemolytic uremic syndrome. If parents choose to give their children unpasteurized juice products, they should do so with caution and be advised that this is an unsafe practice. Commercially prepared unpasteurized juice must contain a warning on the label that the product may contain harmful bacteria.

Research suggests that drinking small amounts of 100 percent fruit juice doesn't affect a child's weight. However, fruit juice contains calories. Just like any other food or calorie-containing drink, too much fruit juice can contribute to gaining of weight. Concerns about the role of fruit juice in tooth decay and childhood obesity, have led to formulation of guidelines for juice intake in babies and children.

To ensure your child isn't drinking too much juice, consider these limits:

Babies younger than 6 months should not drink fruit juice at all. When juice replaces breast milk it can cause nutritional issues. Offering juice before solid foods introducition into the diet replaces breast milk by juice. This can result in reduced intake of protein, fat, vitamins, and minerals such as iron, calcium, and zinc Malnutrition and short stature in children have been associated with excessive consumption of juice .For this age group, breastfeeding is the best..

Babies 6–23 months are still advised to avoid juice completely. It’s more nutritious to serve mashed or pureed fruit instead fruit juice. Never put a baby to bed with a bottle of juice, because it can lead to tooth decay. .

Children 2–5 years should be limited to 125 ml of juice a day. If serving it, give it in a cup along with meals or snacks. Continue to focus on whole fruit over juice .These should preferably be given as fresh juices

Children > 5 years can have 250 ml a day of juice, preferably with meals. These should preferably be given as fresh juices.

What is more beneficial-- whole fruit or fruit juice?

Whole fruit provides you with a whole lot more nutrition that fruit juice. Focusing upon two components of fruit — the skin and the pulp — will help to clarify why there is such a difference between the two.

The benefits of fruit skins

The edible skins of many of the World's Healthiest Fruits - including apples, apricots, blueberries, figs, grapes, pears, plums, raisins, and strawberries - are all sites of important biological activity in the life of the fruit.

The skin is one of the places where the fruit interacts with sunlight, and forms a variety of colored pigments that absorb different wavelengths of light. These pigments, including carotenoids and flavonoids, are well researched as nutrients, that protect our health and nourishment. The skins of whole fruits like grapes have actually been studied for their ability to help lower risk of cancer and help provide protection from ultraviolet light.

Unfortunately, when fruits are juiced, we don't always get to enjoy the fruit's skin.

The benefits of the fruit pulp

In addition to the skin, which is an important source of fibre in most fruits, the pulpy part of the fruit is also a source of fibre (and other nutrients).

Orange juice makes a good example of the health difference when you focus on the issue of its pulp. The white pulpy part of the orange is the primary source of its flavonoids. The juicy orange-colored sections of the orange contain most of its vitamin C. In the body, flavonoids and vitamin C often work together, and support health through their interaction. When the pulpy white part of the orange is removed in the processing of orange juice, the flavonoids in the orange are lost in the process. This loss of flavonoids is one of the many reasons for eating the orange in its whole food form.

Although many commercial products will say "pulp added" on their labels, the "pulp added" many not even be the original pulp found in the whole fruit, and it is highly unlikely to be added back in the amount removed. Juicing reduces the fibre content. Fibre helps normalize bowel movements, lowers cholesterol, helps control blood sugar levels, and aids in achieving healthy weight. Apart from that, it makes you feel full for a longer time (yes much longer. imagine eating a single apple vs. drinking the juice of an apple).

Additionally, many fruit juices that are sold in supermarkets contain only a small percentage of real fruit juice, and contain added sweeteners (sucrose or high fructose

corn syrup). As a result, it is easy to consume a large amount of calories without getting any actual nutrition when you consume these beverages. Make sure you read fruit juice labels carefully! Turnover on the back of the jar or bottle, and look over the ingredient list - you may be surprised to see exactly where the fruit itself fits in!

Conclusions

1. Encourage intake of regional and seasonal whole fruits over fruit juices in children and adolescents
2. Fruit juices/fruit drinks/Sugar Sweetened Beverages should not be offered to infants and young children aged below 2 years
3. For children and adolescents (2-18 years) fruit juices, fruit drinks and SSBs should be avoided as far as possible. Water should be encouraged as the best drink and should be promoted over fruit juices/drinks at home and school.
4. Fruit juices/drinks, if given, should be limited to 125 mL per day for children aged between 2-5 years, and 250 mL per day for age >5 years; and these should preferably be given as fresh juices.
5. Excessive juice consumption may be associated with malnutrition (overnutrition and undernutrition).
6. Excessive juice consumption is associated with diarrhea, flatulence, abdominal distention, and tooth decay. Juice is not appropriate in the treatment of dehydration or the management of diarrhea
7. Unpasteurized juice products may contain pathogens that can cause serious illnesses and should be given to children cautiously..
8. Calcium-fortified juices provide a bioavailable source of calcium and often vitamin D but lack other nutrients present in human milk.

   Pediatricians should support policies that seek to reduce the consumption of fruit juice and promote the consumption of whole fruit by toddlers and young children (eg,child care/preschools).. These should preferably be given as fresh as possible.

Prof. (Dr) Jaydeb Ray
Editor-in-Chief

Spectrum of Childhood vasculitides: Hospital- Based Prospective Data

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Abstract

(a) **Introduction**: Childhood vasculitides are multi-system autoimmune disorders involving different sizes of blood vessel. The relative incidences and the clinical presentations of the different types of vasculitis vary significantly in different geographic area

(b) **Aims and Objectives**: To study the epidemiological pattern, clinical features, etiology and pathological characteristics of childhood vasculitides

(c) **Methodology:** It was an Institution based prospective, observational study at the tertiary care center IPGMER, Kolkata, over a period of 4 years. The patients were diagnosed using ACR (American College of Rheumatology) & CHCC (Chappell Hill Consensus Criteria) criteria. They were undergone relevant investigations and treated according to the standard guidelines. The resultant databases were analyzed.

(d) **Result and Analysis:** A total of 110 patients were diagnosed having vasculitis among them 90% (n=99) were primary vasculitis and the rest are secondary vasculitis (10%, n=11). Majority of the Vasculitis were IgA vasculitis (Henoch Schonlein Purpura) (48.19%,n=53) and Kawasaki disease(31.82%,n=35).They demonstrated various typical and atypical manifestations and a definite seasonal pattern of incidences. Other types of vasculitis include Takayasu arteritis, Poly arteritis nodosa and vasculitis associated with systemic diseases like SLE and JIA. Infection (Scrub typhus) and drug induced vasculitis are also documented. e) Conclusion – A spectrum of vasculitides with a certain trend in their incidences had been delineated from Eastern part of India.

Key Words : Pediatric, Vasculitis, spectrum

Introduction

Childhood vasculitides are multi-system autoimmune disorders involving different sizes of blood vessel. It may be primary or secondary to another illness or a component of many autoimmune diseases. It can range in severity from a self-limited single-organ disorder to a life-threatening disease with the consequence of multiple-organ failure. Secondary vasculitides may be due to drugs, infections or other systemic diseases such as connective tissue disease (CTD) and malignancy[1].

We diagnosed and prospectively followed up

children with different types of vasculitis at our pediatric rheumatology clinic and indoor at the tertiary care center IPGMER, Kolkata, over a period of 4 years from 2015 February to 2019 February. The objective was to study the epidemiological pattern, clinical features, etiology and pathological characteristics in children with vasculitides. An overview of the data obtained is presented here in this article.

Materials and Methods

It was an Institution based, prospective, observational study carried out after obtaining clearance from the institutional Ethics Committee. The study included all consecutive patients between the ages of 1and 12 years attending our hospital with evidence of vasculitis over a period of 4 years. This study was conducted by an integrated approach and support of department of dermatology and pathology. Proper consent and ascent were taken before enrolling the subjects. Patients having any rash due to trauma, insect stings and allergic /urticarial rash (hives) were excluded.

History and clinical examination findings were recorded in a case record form. All clinical data were recorded during the acute presentation of disease. No follow up data was collected as it was a cross sectional observational study. American College of Rheumatology (ACR) and Chappell Hill Consensus Criteria (CHCC) were used for the diagnosis. Digital photographs of the lesion were taken. Routine and specific laboratory investigations were sent according to the requirement for the particular vasculitis syndromes. Those tests as a whole include Laboratory Investigations including complete blood count, Urine for routine examination and microscopy, routine stool examination, liver function test, serum urea, serum creatinine, chest x-ray. HbsAg and Anti HCV, ICTC(Integrated Counseling and Testing Centre for testing HIV), Mantoux (5TU), Antistreptolysin O (nephelometry), Blood Culture (Bactec), Anti Nuclear antibody (Hep20-10,IIFT) , ANCA (Granulocyte, IIFT), Erythrocyte Sedimentation Rate(ESR), C-Reactive Protein (nephelometry), Serum IgG, IgA, IgM (nephelometry), Complement level (nephelometry).Histopathology with Hematoxylin and Eosin staining smear in case of cutaneous vasculitis. Direct Immunofluorescence (where appropriate), Echocardiography, Electrocardiography (where appropriate), Electromyography (EMG), Nerve Conduction Velocity (where appropriate), CT, MRI, MR-angiography, CT-angiography (where appropriate).A punch biopsy (in case of cutaneous vasculitis) was taken from lesions for histopathological examination in 10% buffered formaldehyde solution. Another punch biopsy was taken from lesion skin for direct immunofluorescence (where appropriate) in a suitable transport medium like Michel's transport medium. Data was analyzed by appropriate statistical tests using Statistica version 6. [Tulsa, Oklahoma: StatSoft Inc., 2001]. Data presented as mean for numerical variables and percentages for categorical variables.

Result and Analysis

A total of 110 patients were diagnosed having vasculitis. Among them 64 were male and 46 were female (M:F ratio of 1.4:1). Mean age of presentation was 6.88 ± 2.45 years. Primary vasculitis (90%,n=99) are more common than the secondary vasculitis (10%, n=11).

The distribution of primary Vasculitis were IgA vasculitis (Henoch Schonlein Purpura)

(48.19%,n=53), Kawasaki disease (31.82%,n=35), Takayasu arteritis(2.72%,n=3) and polyarteritis nodosa (PAN)(4.54%,n=5). Secondary vasculitis, constituted of systemic Lupus Erythematosus(4.54%,n=5), systemic Juvenile Idiopathic Arthritis(1.81%,n=2), Urticarial vasculitis(1.81%,n=2), Anti-thymocyte globuline induced arthus reaction(0.09%,n=1),and penicilamine induced Anti-neutrophilic cytoplasmic antibody(ANCA) positive vasculitis (0.09%,n=1), infection (scrub typhus) induced vasculitis(1.81%,n=2).Interesting to note a total of 88 cases out of 110 cases (80%) constitute either HSP and or KD. All other cases constitute only one fifth fraction of the population indicative of the rarity of the vasculitis other than HSP and KD.

Table 1: Different types of Vasculitis according to the ACR and CHCC classification

Vasculitides (N=110)	Number of cases	Frequency (%)
Large Vessel Vasculitis		
Takayasu arteritis	3	2.72
Medium Vessel Vasculitis		
Kawasaki disease	35	31.82
Polyarteritis Nodosa	5	4.54
Small Vessel Vasculitis		
IgA vasculitis (Henoch-Schönlein)	53	48.19
Hypocomplementemic urticarial vasculitis	2	1.81
Anti–glomerular basement membrane disease	1	0.09
Vasculitis associated with systemic disease		
SLE	5	4.54
JIA	2	1.81
Vasculitis associated with probable etiology		
Infection (Scrub Typhus)	2	1.81
Penicilamine induced ANCA positive vasculitis	1	0.09
ATG induced arthus reaction	1	0.09

Major primary Vasculitis was HSP (48.19%,n=53). Mean age of presentation of HSP was 7.5 yrs (range 5-12 yrs) with female preponderance (M:F= 1:1.45). All had Palpable Purpura (100%,n=53). Gastro-Intestinal involvement was noted in 44 patients (83%,n=44) .Other typical features were noted in variable frequencies. Four patients had orchitis and 2 had convulsion. One child presented with recurrence after 3 months of initial episode. Among the uncommon features notable were raised Anti-streptolysin O(ASO)titre(n=10), low C3 without any renal involvement(n=4), raised serum IGA(n=8). Histo-Pathological Examination (done in 45 patients) revealed Perivascular lymphocyte predominant infiltrate (88.9%,n=40) in majority. Direct Immunofluroscence (DIF) could be done in 28 patients. Among them 8 were DIF negative and 20 were DIF positive. All patients of DIF positivity had shown 100% positivity for Ig A, C3,Fibrin deposition.2 cases had shown IgM positivity.

Kawasaki disease (KD), the second commonest Vasculitis (31.82%,n=35), with mean age 5.3 years, (M:F=4:3) showed all the typical presentations in majority. The diagnosis of incomplete KD was recorded in 9 patients. Atypical features include Peri-anal desquamation (25.7%,n=9), diarrhea (20%,n=7),arthritis(11.4%, n=4). Four patients developed HLH (11.4%,n=4) of which 1 of them had dengue. One patient presented with recurrence who had been diagnosed as incomplete Kawasaki disease 2 years back. Coronary Artery Disease (CAD) was detected on echocardiography in 7 patients (20%.The coronary artery involvement showed the involvement of Left Main Coronary Artery involvement-4 (57.1%), Left Anterior

descending coronary artery involvement- 1 (14.3%),Circumflex coronary artery- 1 14.3%), Right Main Coronary Artery -1 14.3%).

All of the 3 patients with Takayasu disease were female and mean age of presentation was 7.7 yrs. All had Descending thoracic aorta involvement along with systemic hypertension. Five patients had PAN (M:F ratio 3:2, mean age of onset - 5.5 yrs) of which 1 had cutaneous PAN. All had fever, myalgia, and Skin involvement. SLE associated vasculitis was found in 5 cases. One of them had CNS vasculitis. Over the last three years incidence of scrub typhus had been increased in the eastern India. We have two patients with necrotizing vasculitis and gangrene.

Seasonal pattern of Vasculitis has been observed in our study. The comparison of HSP and KD, the major vasculitides has been presented here. Incidence of HSP was common in autumn and that of KD was in the winter months. The incidences of overall cases were less during the monsoon.

Table 2: Seasonal Trend of major vasculitides HSP and KD

Season	HSP (n=53) (%)	KD (n=35) (%)
Winter	10 (18.9)	17 (48.5)
Autumn	24 (45.3)	8 (22.9)
Summer	14 (26.4)	8 (22.9)
Monsoon	5 (9.4)	2 (5.7)

Discussion

The term vasculitis (vasculitides, pleural) indicates the presence of inflammation in a blood vessel wall. The inflammatory infiltrate may be predominantly neutrophilic, eosinophilic, or mononuclear. Perivasculitis describes inflammation around the blood vessel wall but without mural involvement. Vasculopathy, a broader term, indicates an abnormality of blood vessels which may be inflammatory, degenerative, or may result from intimal proliferation[1].

The vasculitides are the most difficult of all rheumatic diseases to classify. Traditionally, vasculitis syndromes had been categorized according to features that include clinical phenotype, the predominant size of the involved vessels, or the histopathology of the involved vessel. In 1990 a committee of the American College of Rheumatology (ACR) provided formal classification criteria for many, but not all, individual types of vasculitis[2]. In 2012, the International Chapel Hill Consensus Conference (CHCC 2012) updated the 1994 consensus recommendations (CHCC 1994) on the names of diseases, preferred abbreviations, and disease definitions[3,4]. CHCC 2012 retained the ACR framework for categories of vasculitis based on the size of predominantly affected blood vessels defined as follows.

In our study, the most common vasculitis found is HSP (48.19%,n=53), followed by KD (31.82%,n=35).This pattern is similar to western and Indian data[5-8]. Majority of the study in pediatric vasculitides are available from western countries[6-8]. However, there are only hand full of pediatric study available from Indian perspective[5].

In our previous published data of seven years (2004-2011), out of 158 cases of vasculitides, HSP was the major vasculitis (56.9%,n=90) followed by KD (24%,n=38)[5].Our present study is also showing the same trend. The United States series of 434 patients showed 213 patients with HSP (49.1%) and 97 patients with KD (22.4%)[6]. Data from Turkey as published by Ozen et al. in their series of 376 patients also showed majority with HSP

than KD (218 vs.78)[8]. In the Canadian series, KD was shown to be more frequent than HSP[7]. The incidence of diagnosis of KD in India has apparently increased over past decades due increase in awareness, however HSP is still outnumbering the KD in this part of the country.

Among the subjects of HSP, the mean age at presentation was 7.5 years which is comparable to that reported earlier[9]. Skin involvement occurred in all patients that included purpuric rash (100%) and subcutaneous edema (48.1%). Abdominal pain preceded the rash in 6 (22.2%) patients. Arthritis, angioedema was initial presentation in 2 patients each. Various Clinical features of our HSP cohort is comparable to other study cohort is described below in tabular form[10-12] Neurological manifestations are also unusual accompaniment of HSP and include coma, subarachnoid hemorrhage, seizures and Guillain-Barre Syndrome[9]. We had one child presented only with fever and convulsion, later developed rash and the other one developed convulsion on day 3 of illness.Treatment given was supportive with maintenance of good hydration, nutrition, and electrolyte balance. Control of pain is accomplished with simple analgesics in 38 patients, antihistaminic was given to 48 children. Oral Prednisolone was given in 10 children while IV methyl prednisolone was given to 2 cases. Anti-hypertensive was prescribed in 5 children. With standard treatment all patients achieved remission with mean disease duration of 14.7 days. Recurrence is uncommon in HSP and was noted in 1 child after 3 months.

KD is the second most common vasculitis in our study population (31.82%, n=35). Although it is classically described as disease of younger children (<5 years), our study showed more older children (>5 years) with male preponderance similar to various other part of India[13,14]. In our case series majority had classical KD (74.3%,n=26) and 9 cases were incomplete KD((25.7%, n=9)). Fever, rash, and nonexudative conjunctivitis, peri-ungual desquamation were present in all patients. CAD was found in seven patients and the Incidence of CAD is similar to Chandigarh data[14]. Four of our study cohort developed HLH (11.1%, n=2) of which 1 of them had dengue, the data of which has already been published in our series[15]. Recurrent KD in India have been rarely reported[16]. One patient of our study cohort , who was diagnosed earlier as incomplete KD, had recurrence after two years. The case has been published as the first case of recurrent Kawasaki from eastern India[17]. All patients were treated initially with IVIG.IVIG resistance was noted in 3 of our patients who responded thereafter. No one in our series received infliximab. Dexamethasone was given in patients with HLH.

All of the 3 patients with Takayasu disease in our study cohort were female and mean age of presentation was 7.7 yrs. Age of presentation was less compared to other Indian study[18]. It occurs more frequently in adolescence than in childhood, but it can occur in very young children. All of the patients had hypertension at initial clinical presentation. Echocardiography revealed Left ventricular dysfunction in 2 patients. All had Descending thoracic aorta involvement in angiography. These clinical finding is comparable to previous Indian studies[18]. All received Prednisolone and Antihypertensive, while methotrexate was given in one child.

Five of our patients had PAN (M:F ratio 2:1, mean age of onset - 5.5 years) of which 1 had cutaneous PAN. All had fever, myalgia, and Skin involvement. Diagnosis was established in all patients by skin biopsy which showed necrotizing vasculitis of the medium sized vessels. None had renal or pulmonary involvement. Our findings are comparable to our previous literature from eastern India[19].

Vasculitides secondary to systemic disease and drugs are not uncommon[20]. Histopathological confirmed seven of our case had vasculitis secondary to systemic disease; five were diagnosed as SLE while two other as sJIA. Two of our study cohort was histopathologically diagnosed as a case of urticarial vasculitis and one patient with renal failure was diagnosed having anti GBM disease. Drug associated vasculitides was noted in two children. Anti-thymocyte globulin was given to one patient of aplastic anemia which caused Arthus reaction (type III hypersensitivity reaction). One of our children with Wilson's disease was diagnosed as penicillamine-induced ANCA positive vasculitis. Penicillamine causing such vasculitides has been rarely reported[21]. Scrub typhus infection associated vasculitis had been found in 2 cases one of which developed pan-digital gangrene. Focal or disseminated vasculitis caused by the endothelial cell destruction and the perivascular leucocytic infiltrations are the main pathological changes occur[22].

Conclusion

The childhood vasculitides are not uncommon in this part of the world. A certain trend has been found where Henoch Schonlein Purpura remained as the predominant cause of vasculitis in children followed by Kawasaki Disease and collectively they form the majority of cases. They respond well to the treatment if the diagnosis is made early and there may be significant modification in terms of morbidity and mortality. However multicentric studies involving large number of subjects over a longer period are needed for proper delineation of the spectrum of childhood vasculitis.

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Critical Monitoring and General Care in PICU – A Practical Approach

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Introduction:Intensive care involves the monitoring and support of multiple organ systems tailored to the individual patient. The success of all intensive care areas depends onearly identification of a critically sick child followed by prompt decision making by the caregivers for early institution of appropriate and rational treatment andoverall supportive careprescribed for the particular child. Theaim in intensive care management is to target the primary or precipitating disorder but to provide holistic care to a sick child due attention should be rendered to supportive care with equal importance.All sick children need regular monitoring which can be technology based and clinical. But as a caregiver, everyone should remember that the technology only adds to the clinical monitoring but cannot replace clinical monitoring[1].

What are the purposes of monitoring?

The three main purposes of monitoring are as follows:

(a) To measure key physiologic indices that help in diagnosis and management.

(b) To provide guidance to the health care team regarding the important changesthose have occurred in the child's condition.

(c) To identify and evaluate trends that would help in the assessment of treatment and prognosis of the patient[1].

All data should be recorded on a pre-designed simplified chart. The recent evidence based guidelines should be followed in all aspects. This is a genuine fact that the intensive care is always dynamic which involves evaluation-intervention-re-evaluation.

How to assess a child in PICU?

Each patient in PICU needs to be completely examined and assessed at least twice a day and more often if unstable. A large amount of information will be available from the charts, monitors and from clinical examination. It is important to have a uniform system for condensing the information easily so that patient assessment can be made, plan formulated and communication between different teams and shifts facilitated. An example is given below in relation to our CVS: To formulate a check list so that caregivers

will maintain uniformity all throughout the PICU stay[3].

(a) To monitor HR (trends and actual values), BP (check if age appropriate), CVP if available. Check doses of inotropes (if any).

(b) To look for adequacy of systemic perfusion: CRT, peripheral pulse volume, peripheries (cold/sweaty/warm).

(c) To look for evidence of adequate organ perfusion: Consciousness level, urine output, metabolic acidosis.

(d) To examine all IV lines : whether it is clean or has evidence of infection.

The dictum of"First Do No Harm" must be followed by the entire health care team. Due attention should be focussed on homeostasis, fluid and electrolyte balance, temperature control, nutrition, sedation and analgesia along with prevention and control of infection.

Blood glucose control:

In all children admitted in PICU the caregivers should try to prevent hypoglycemia (< 50 mg %) to reduce morbidity and to target normoglycemia(80-110 mg %) (4). The normal blood sugar levels protects CVS, prevents from secondary infections.It is already documented that the short- term hyperglycemia in initial phase may be helpful but prolonged hyperglycemia is detrimental. If blood glucose levels are consistently more than 180 mg% despite reduction of glucose infusion rates, initiating insulin infusion may be considered. It is recommended to monitor blood glucose regularly and to target to keep the blood sugar levels below 150 mg % which is attainable and realistic.

Pressure sores:

Pressure sores are also reported in children admitted in PICU. The children at high risk are children on mechanical ventilator, children with hypotension. Other risk factors are malnutrition, sensory loss, dependent edema, and PICU stay more than 96 hours. It is very essential to examine the skin regularly in all PICU patients.The preventive strategies recommended are to maintain adequate nutrition, use of alternating pressure mattresses(Fig: 1),position change and re-positioning frequently depending upon the results of skin inspection.

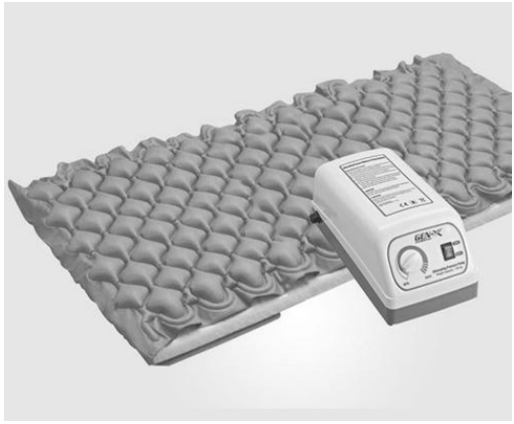

Fig: 1- Pressure mattress

Eye care of all sick children:

Corneal and conjunctivaldryness, erosion are not uncommon in PICU patients. The patient'sinability to fully close the eyes, use of NM blocking agents are the proven risk factors. It is advised to use protective eyelid taping and to use of lubricating ointments and artificial tears to prevent conjunctival and corneal complications in PICU.

Oral hygiene in PICU:

Oral and dental care is one of the most

commonly neglected issues in most of the PICUs all over the world. But oropharyngeal colonization is of significant importance in all sick children. Prevention of colonization may be an effective method of infection control. Plaque accumulation and gingival inflammation are seen in critically sick children. All critically ill children are at risk from local and systemic spread of oral microorganisms. It is recommended to do regular cleaning of oral cavity especially in intubated patients.Use of chlorhexidine based oral hygiene products are helpful[1].

Prevention of Stress Ulcers:

PICU patients are at risk of developing stress ulcers in the stomach. There are few definite risk factors for upper GI bleeding in critically ill children. These arethrombocytopenia, prolongedaPTT,children on mechanical ventilation, very severe illnesses and children on steroid therapy.The stress ulcer prophylaxis in children can be started withH2 blockers, proton-pump inhibitors and oral sucralfate.

Thromboembolic prophylaxis:

Venous thrombosis is being increasingly recognized in the pediatric age group. The important risk factors are -underlying diseases, steroid therapy and selected therapeutic intervention like the insertion of central venous catheter in femoral veins.Standard unfractionatedheparin remains the mainstay of therapy and prophylaxis; but now LMW heparins are being increasingly used.Also benefit of prophylaxis must be weighed against the risk of bleeding complications.DVT prophylaxis in post-pubertal children with sepsis is recommended in PICU.

Care of IV lines, central lines, chest tubes:

IV cannula must be examined at least once per shift for signs of infusion phlebitis.Visual Infusion Phlebitis (VIP) scale can be used routinely for objective assessment.Central venous and arterial lines should be monitored regularly for displacement, bleeding, patency, infection or hematoma formation.

They should be removed as soon as they are no longer necessary. The ICD tubes should be assessed periodically for movement of fluid column, amount of drainage and its nature,displacement, breath sounds any evidence of subcutaneous emphysema.

Care of children with special needs:

Children with neurologic and neuromuscular disorders have special needs.After extubation they are in need of prolonged support in the form of nasal CPAP or tracheostomy.They require special attention on nutritional support, regular changing of positions and physiotherapy for prevention of contractures and pressure sores.

Nutritional support of PICU patients:

Malnutrition increases complications and worsens outcomes for critically ill patients. The problem of under-nutrition is compounded by the decreased intake, accelerated demands brought by acute phase of the illness.As the children have less energy and substrate reserves they are more dependent on substrate supply than adults.It is essential to provide calories in accordance with REE-resting energy expenditure which is more physiological.In initial unstable resuscitative phase 'permissive hypocaloric nutritional

support' is recommended @ 20-30 Kcal/Kg/day. It is recommended to switch on to 'hypercaloric support' once the recovery phase sets in.

Enteral route nutrition:

The oral route is the preferred one provided the GIT is functioning. This route is safer and cost-effective also.Enteral nutrition helps to maintain gut barrier; reduces the risk of bacteremia and pneumonia.The major contraindications of oral route are severe GI hemorrhage, recent GI surgery, NEC, severe vomiting or diarrhea.

Parenteral route nutrition:

Parenteral nutrition is delivery of all the nutrients directly to the blood stream.

Glucose infusions are started @ 5-6 mg/Kg/min; amino acids @ 1 gm/Kg/day, then it are increased over 2-3 days to 2.5 gm/Kg/day.Lipids are infused @ 0.5 gm/Kg/day,and then it is increased to 2.5 gm/Kg/day over 4-5 days. Appropriate combinations can be achieved on the basis of the fluid requirements. But this route is associated with many inherent complications; needs a regular bio-chemical monitoring in all patients.

Sedation, analgesia and paralysis:

Goal of sedation is safe and effective control of pain, anxiety and stress so as

to allow a necessary procedure to be performed andto provide appropriate amnesia or decreased awareness.Pre-sedation evaluation along with careful monitoring during and after procedure is the key to prevent complications.NMBDs must always be administered after adequate sedation has been achieved as these drugs don't have any sedative or analgesic effect.Children receiving NMBDs should simultaneously receive sedative and analgesic for overcoming anxiety and pain[1].

Control of nosocomial infection:

All areas offering intensive care should have a definite 'infection control program'.It reduces nosocomial infection rates by up to 50 %. Training of nursing staff and attending physicians is essential for infection control. Proper hand hygiene(hand washing with soap and water or by using alcohol based anti-septic solution), disinfection of instruments (resuscitation bags, ventilator circuit, nebulizers), positioning of patients with head end elevation (to reduce the risk of aspiration) and strict aseptic precautions should be followed whenever doing an invasive procedure ; they will ultimately help to reduce nosocomial infection. (Fig: 2)

What is FAST HUG?

FAST HUG is a well-known mnemonic that stands for Feeding, Analgesia, Sedation, Thromboembolic prophylaxis, Head-of-bed elevation, Ulcer (stress) prevention, and Glucose control. This is a simple mnemonic to highlight SEVEN key aspects in general care of all critically ill patients.This should be

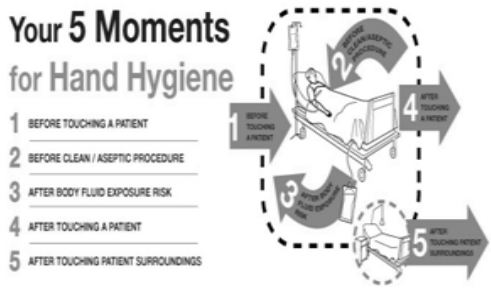

Fig: 2- WHO 5 moments of hand hygiene

considered at least once a day during rounds and, ideally, every time the patient is seen by any member of the health care team.It can be applied to every ICU patient, as it is not restricted to any specific group. The mnemonic is long enough to include fundamental aspects of care that involve all members of the care team but short enough to be easily remembered.It has a personal touch- as we all like a hug, and our patients are no exception[2].

Conclusion:

Intensive care is built up on the foundation of monitoring and supportive care. Rationalcare should be provided with due compassion and concern. Critical care is highly labor-intensive and this always demands a good quality team work.

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Quiz Answer

Netherton Syndrome. Genetic analysis will reveal mutations in the *SPINK 5* gene, which encodes a serine protease inhibitor(*LEKT1*).

Madhumita Nandi
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(Due permission from parents taken for publication of the picture)

Management of Sero Discordant Couples of HIV

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NACO Launched Test and Treat Strategy on 28th April 2017 irrespective of CD4 Count, age or WHO Clinical Staging.

Couple is defined as Two persons in an ongoing sexual relationship; each of these persons is referred to as a "partner" in the relationship.

A serodiscordant couple is a couple in which one partner is HIV-positive and one partner is HIV-negative. Although one partner is currently HIV-negative, this does not mean that this partner is "immunized" or protected against getting HIV in the future.

To date, HIV testing and counselling approaches have largely focused on individuals, despite the fact that many people in stable relationships and marriages get infected with HIV.

Many men and women in stable relationships are unaware of their partner's HIV status, and many people with an HIV-positive partner are not aware of their own status. Up to fifty percent of people living with HIV who are in relationships are estimated to be part of discordant couples,

It is of paramount importance for serodiscordant couples to avoid transmission to the HIV-negative partner. It is possible for couples to stay HIV serodiscordant indefinitely if they consistently practice safer sex using male and female condoms.

The annual risk of transmission of HIV from an infected partner to an uninfected partner in serodiscordant couples can be reduced from 20–25% to 3–7% in programmes where condom use is recommended for prevention.

The HIV-positive partner should receive care and treatment services for his or her own

health. Treatment for the HIV-positive partner also is highly effective in reducing the risk

Outcomes that occur:

- Infection of the non-infected spouse
- Re-infection for both of them
- Abandonment of the positive partner especially in case of dependence, e.g. a housewife who is dependent on her husband

Use of PrEP by serodiscordant couples[1]

In serodiscordant couples efforts to prevent HIV/STI should first and foremost follow the recommendations set forth in Guidance on couples HIV testing and counselling, including antiretroviral therapy for treatment and prevention in serodiscordant couples .This guidance recommends the use of early

treatment with antiretrovirals for the infected partner to reduce chances of HIV transmission. Countries should decide what to recommend to serodiscordant couples: early initiation of treatment for the infected partner, PrEP for the uninfected partner, or a combination of the two. Best approaches will likely vary across contexts and may need to be tailored to specific situations.

Recommendation

:In countries where HIV transmission occurs among serodiscordant couples, where discordant couples can be identified and where additional HIV prevention choices for them are needed, daily oral PrEP (specifically tenofovir or the combination of tenofovir and emtricitabine) may be considered as a possible additional intervention for the uninfected partner.Conditional recommendation, high quality of evidence.

ART for prevention in serodiscordant couples

People living with HIV who are in serodiscordant couples and who are started on ART for their own health should be advised that ART is also recommended to reduce HIV transmission to their uninfected partner. Strong recommendation, high-quality evidence.

Antiretroviral therapy for HIV-positive partners with >350 CD4 cells/μL in serodiscordant couples should be offered to reduce HIV transmission to uninfected partners. Strong recommendation, high-quality evidence.

The HPTN 052 randomized controlled trial found a 96% reduction in HIV transmission in serodiscordant couples where the partner with HIV with a CD4 count between 350 and 500 cells/μL had started ART early.

Children in Discordant Couple

Before attempting conception, the partner living with HIV should be on ART and have achieved sustained suppression of plasma viral load below the limits of detection.

It is important to recognize that no single method (including treatment of the partner living with HIV) is fully protective against transmission of HIV, though the risk appears to approach zero when the partner living with HIV maintains a consistently undetectable plasma viral load on ART.

In addition to reducing the risk of HIV transmission between partners, starting ART before conception in women living with HIV may also further reduce the risk of perinatal transmission.18Evidence suggests that early and sustained control of HIV may decrease the risk of perinatal transmission,19,20 but it does not completely eliminate the risk of perinatal transmission.

1. Assisted insemination via sperm washing technique (Husband reactive)+ wife (non reactive) should be on PrEP
2. When a man living with HIV is in a serodiscordant relationship, the use of donor sperm from a man without HIV is an option for conception that eliminates the risk of HIV transmission to the partner without HIV.
3. Attempt conception (sexual intercourse without a condom limited to peak fertility) with wife reactive + Husband (non reactive) should be on PrEP
4. Adoption

Pre-exposure Prophylaxis (PrEP)

- Pre-exposure prophylaxis (PrEP) is an HIV prevention strategy where HIV-negative individuals but who are at substantial risk of getting it take anti-HIV medications before coming into contact with HIV to reduce their risk of becoming infected. The medications work to prevent HIV from establishing infection inside the body.

- PrEP has been shown to reduce risk of HIV infection through sex for gay and bisexual men, transgender women, and heterosexual men and women, as well as among people who inject drugs.
- It does not protect against other sexually transmitted infections (STI) or pregnancy. It is not a cure for HIV

The ultimate goal of PrEP is to reduce the acquisition of HIV infection with its resulting morbidity, mortality, and cost to individuals and society

The principle of PrEP is –

Somebody who does not have HIV takes enough ARVs for there to be high levels of the drugs in their bloodstream, genital tract and rectum before any exposure to HIV.

- If exposure occurs, the ARVs stop the virus from entering cells and replicating.
- This prevents HIV from establishing itself and the person remains HIV negative.
- The antiretrovirals which are currently used as PrEP is a combination of two drugs (tenofovir and emtricitabine).

These ARVs were chosen because they have limited side-effects, have few problems with drug resistance, reach high levels in the genital tract and rectum, and remain in the body for a relatively long time.

Eligibility criteria include:

- HIV-negative
- no suspicion of acute HIV infection
- substantial risk of HIV infection
- no contraindications to PrEP medicines (e.g. TDF/FTC)
- willingness to use PrEP as prescribed, including periodic HIV testing
- Contraindications to PrEP
- HIV infection
- signs/symptoms of acute HIV infection, probable recent exposure to HIV
- estimated creatinine clearance of less than 60 ml/min (if known)
- allergy or contraindication to any medicine in the PrEP regimen
- Medications approved for PrEP
- TRUVADA (TDF/FTC)
- The U.S. Food and Drug Administration (FDA) approved Truvada for PrEP use in 2012.
- This medication is taken as a once-daily oral pill, which combines two medicines in one: and Tenofovir disoproxil fumarate or TDF (300 mg) and Emtricitabine or FTC (200 mg)
- Truvada works by blocking an enzyme called HIV reverse transcriptase.
- By blocking this enzyme, it prevents HIV from making more copies of itself in the body.
- Side Effects of PrEP
- Nausea: Headache:
- Diarrhoea :
- liver health :
- kidney health :

Clinical Follow-up and Monitoring

- At least every 3 months to
- Repeat HIV testing and assess for signs or symptoms of acute infection to document that patients are still HIV negative
- Repeat pregnancy testing for women who may become pregnant
- Provide a prescription or refill authorization of daily TDF/FTC for no more than 90 days (until the next HIV test)
- Assess side effects, adherence, and HIV acquisition risk behaviors

- Provide support for medication adherence and risk-reduction behaviors
- Respond to new questions and provide any new information about PrEP use
- Conduct STI testing for sexually active persons with signs or symptoms of infection
- At least every 6 months to
- Monitor CrCl
- If other threats to renal safety are present (e.g., hypertension, diabetes), renal function may require more frequent monitoring or may need to include additional tests (e.g., urinalysis for proteinuria)
- A rise in serum creatinine is not a reason to withhold treatment if CrCl remains =60 ml/min.
- If CrCl is declining steadily (but still =60 ml/min), consultation with a nephrologist or other evaluation of possible threats to renal health may be indicated.
- Conduct STI screening for sexually active adolescents and adults even if asymptomatic
- At least every 12 months to
- Evaluate the need to continue PrEP as a component of HIV prevention

Acknowledgements

- WHO- Guidelines and Publications on Pre -Exposure Prophylaxis (PrEP)
- CDC - Guidelines and Publications on Pre -Exposure Prophylaxis (PrEP)

Announcement

Kawasaki Disease Mimicking Retropharyngeal Abscess – An Unusual Presentation – A Case Report

Souradeep Chakrabarti*, Shirsendu Dutta*, Rohit Bannerji, Jaydeb Ray*****

Post graduate trainee, ** Associate professor, * Professor and Head of the Department, Department of Pediatrics, Institute of Child Health, Kolkata.*

Abstract: Kawasaki disease ,formerly known as mucocutaneous lymph node syndrome is an acute febrile illness of childhood. It is a medium vessel vasculitis can affect multiple organs and systems. Retropharyngeal cellulitis is a rare but known manifestation of Kawasaki disease. Here we are presenting a case of a 1 year 8 month old boy who presented with high grade fever for 3 days with soft tissue swelling of left side of the neck with restricted movement mimicking an acute retropharyngeal abscess. Initially antibiotics were started but subsequently the child developed other signs and symptoms of Kawasaki disease. The child was successfully treated with one dose of intravenous immunoglobulin (IVIg) and aspirin.

Keywords: Kawasaki disease, retropharyngeal cellulitis, vasculitis, IVIg

Introduction

Kawasaki disease is a vasculitis that predominantly affects medium sized arteries. The coronary arteries are most commonly involved. It has been increasingly recognized that cervical lymphadenopathy can be associated with deep neck inflammation leading to parapharyngeal and retropharyngeal edema and non suppurative phlegmon[1,2].This non suppurative retropharyngeal phlegmon is often misdiagnosed as acute retropharyngeal abscess and sometimes requires unnecessary antibiotics use and surgery.

Case report

A 1 year 8 month old boy presented with high grade fever for 2 days associated with left sided neck swelling (Fig:1) and extreme irritability. Swelling was warm and tender. There was gradual restriction of neck movement. On examination there was a lymph node at left sided posterior triangle,

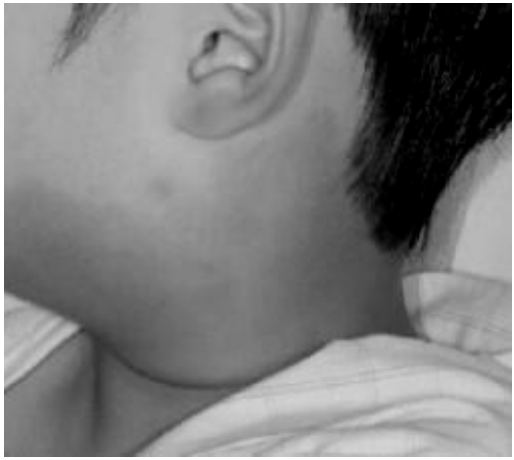

Fig 1. Left sided neck swelling mimicking retropharyngeal abscess.,

(4x3.5) cm2 in size and freely mobile. Liver was palpable 3 cm below costal margin. Lips were erythematous. Suspecting a case of acute retropharyngeal abscess IV antibiotics (Flucloxacillin and Vancomycin) were started empirically after sending routine blood investigations including blood culture. Routine blood examinations revealed hemoglobin 11.1 g/dl, Total leukocyte count was 18600/cmm (Neutrophil 63%,Lymphocyte 26%).Platelet count was 4.6 lac/cmm. C reactive protein was 64mg/lit (Normal<6mg/lit). Erythrocyte sedimentation rate was 89mm/h(Normal <15 mm in first hour). Serum urea, creatinine, electrolytes, liver function tests all were within normal limit. Urine routine and microscopic examination revealed 15-20 pus cells but urine culture was sterile. In spite of giving IV antibiotics fever spikes increased and the child became more toxic in next 24 hours. MRI of neck was done and it revealed left sided retropharyngeal edema, cellulitis and phlegmon.(Fig:2). But 2 days after hospital admission, on day 4 of fever the child gradually developed increased erythema and cracking of lips,strawberry tongue, bilateral non purulent bulbar conjunctival injection and erythema and edema of hands and feet .There was also perianal excoriation. There was no rash in the body. Thus the child was diagnosed as a case of Kawasaki disease based on the clinical criteria. The child was treated with a single dose of intravenous immunoglobulin(IVIg) (2g/ kg over 12 hours) along with aspirin (50mg/ kg/day). Fever and conjunctival injection subsided within 24 hours after the completion of IVIg treatment, whereas edema of palms and soles and neck swelling resolved gradually within next 48 hours after IVIg treatment. Blood Culture was sterile, so antibiotics were stopped after 4 days of use and all the signs of retropharyngeal involvement resolved without any further treatment. Echocardiography showed no coronary artery involvement. Complete hemogram and CRP were repeated 48 hours after subsidence of fever. Hemoglobin was 11.4g/dl, total leukocyte count was 12600/cmm (Neutrophil 50%,Lymphocyte 36%), platelet count was 7.9lac/cmm and CRP was 4.6mg/lit. Dose of aspirin was reduced to 5mg/kg/day. Desquamation of the fingertips started from the day 10 illness. The child was discharged from the hospital with no sequelae on the 10th day of illness. Echocardiography was repeated 2 weeks after the first echocardiography and it did not reveal any abnormality. Aspirin was stopped after 6 weeks. A 6 month follow up revealed no remarkable additional morbidity and complication regarding with Kawasaki disease.

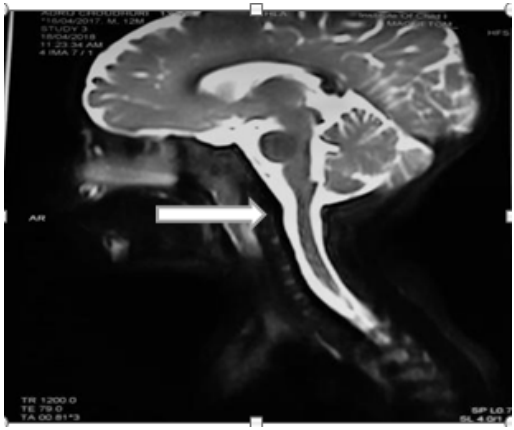

Fig 2. MRI of neck showing retropharyngeal edema, cellulitis and phlegmon.

Discussion

Kawasaki disease (KD) is a self limiting vasculitis mainly involves medium size arteries. For the majority of patients Kawasaki disease is a disease of early childhood, mainly occur in Asian children and nearly all epidemiologic study shows a higher susceptibility to KD in boys. The coronary ateries are most commonly involved, although

other arteries, such as the popliteal and brachial arteries can also develop dilatation. The life threatening complications of Kawasaki disease includes coronary artery aneurysm, shock and macrophage activation syndrome. Kawasaki disease is always a clinical diagnosis. The classic clinical criteria includes persistent unremitting fever (38.30C) for 5 days and unresponsive to antibiotics with presence of at least 4out of 5 principal features:

1. Erythema of palms and soles and edema of hands and feet in acute stage and in subacute stage periungual peeling of fingers and toes from second week of illness.
2. Polymorphous exanthema.
3. Bilateral non purulent bulbar conjunctival injection,
4. Erythema and cracking of lips, strawberry tongue and diffuse injection of oral and pharyngeal mucosa.
5. Cervical lymphadenopathy (>1.5 cm in diameter) usually unilateral. Laboratory findings in acute Kawasaki disease include: neutrophilic leukocytosis, elevated ESR and C reactive protein,anaemia, hypoalbumine-mia, hyponatremia,abnormal plasma lipid, sterile pyuria, elevated serum transaminases, elevated gamma glutamyl transpeptidase and occasionally pleocytosis in CSF[3].

Retropharyngeal edema which often mimics retropharyngeal abscess is a rare complication of Kawasaki disease with various differential diagnoses, including infection in the retropharyngeal space, trauma, neoplasm, post radiation therapy, acute longus coli tendinitis and crown dense syndrome[4]. In the present case ,all other previously known causes of retropharyngeal edema were ruled out and this condition was attributed to KD. A review of literature indicated that this complication which is generally diagnosed based on CT findings, was first described by Pontell et al. In 1994[5]. Subsequently many case reports have been published. In a study of Kanegaye et al, 12 patients with KD who presented with lymphadenopathy were evaluated retrospectively, eleven of them had neck CT scan done and none of them showed abscess formation. Retropharyngeal oedema was found in 7 of these cases[1]. Fang et al reported a case of KD (3-year old girl) presenting with retropharyngeal edema and shock syndrome who had an open surgical exploration and culture was found to be sterile[6]. A case was reported by Kao *et al*, extensive involvement in retropharyngeal space and airway compression regressed after IVIg treatment on repeat imaging[7]. The pathophysiology of KD associated retropharyngeal edema is unknown. However, inflammation and edema associated with systemic vasculitis and increased microvascular permeability have been suggested as primary mechanisms[8].

In the present case the diagnosis of Kawasaki disease was delayed, preventing us from administering intravenous immunoglobulin (IVIg) in proper time. As in the present case, early therapeutic intervention is sometimes difficult because there are no specific diagnostic test for Kawasaki disease. Diagnosis is entirely based on clinical criteria. Kawasaki disease should be kept in mind if a child presents with fever with symptoms mimicking retropharygeal abscess not responding to antibiotic therapy especially when associated with other signs and symptoms of Kawasaki disease. Thus with a prompt diagnosis and treatment with IVIg and aspirin coronary complications and unnecessary antibiotic use and surgical drainage could be avoided.

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IAP Odisha Activity

Asthma Training Module Workshop
Venue: All India Institute of Medical Sciences, Bhubaneswar on 10th March 2019.

IAP under Graduate Quiz (Division round)
Venue: AIIMS, Bhubaneswar on 24th August 2019.

Zonal Infectious Diseases CME
Venue: IMA House, Bhubaneswar, on18th August 2019

IAP ALS Provider Course
Venue: AIIMS Bhubaneswar, **Date:** 30th & 31st March 2019

Salient Features of The Updated Pediatric TB Guidelines 2019

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The guidelines on Pediatric TB have been revised by Indian Academy of Pediatrics & Revised National Tuberculosis Control Program in February 2019. This is the first revision of the guidelines that were published and recommended in 2015. The updated guidelines are an effort to fast track the use of newer diagnostics and therapy modifications for managing TB in children. There are several salient changes in these guidelines that are being highlighted here.

1. Relevant symptomology has a new criterion -

In the criteria for no weight gain or weight loss, it has now been added to consider "Poor weight gain despite nutritional rehabilitation in children with SAM" as one of the criteria of poor weight gain. The following set of symptoms are to be considered for suspecting TB in a child –

- Persistent fever for 2 weeks or more, without a known cause and/or
- Unremitting cough for 2weeks or more and/or
- Weight loss of 5% in 3 months or no weight gain in past 3 months
- Poor weight gain despite nutritional rehabilitation in children with SAM
- Contact with a patient with Pulmonary TB in past 2 years

2. When to do CBNAAT?

In the interim guidelines of 2016, it was recommended that CBNAAT should be done upfront in all children with suspected TB even before doing a Chest X Ray.

The current recommendations state that achild with suspected pulmonary TB should have a Chest XRay done first along with a skin test for TB. If the Chest X Ray is highly suggestive of tuberculosis, then sputum/ gastric lavage/ induced sputum sample should be subjected to a WRDT (WHO approved Rapid Diagnostic test). Therefore the presence of symptoms along with suggestive radiology should guide the ordering of rapid diagnostic tests such as CBNAAT.

The rationale for this recommendation is that a lot of over ordering of CBNAAT was being done which showed positive results in only 10% of the samples that were being subjected to the test. With the new recommendations, the yield is expected to jump to 30-40%.

3. Skin test for TB

TST (Tuberculin skin test) which till date is

the Mantoux test in India faces a few issues. The source lot of tuberculin is finished and equivalent lots may not be the same and generate equivalent results.

Newer generation tests are likely to replace the tuberculin skin test. Of all such tests, the C-TB test is showing promise and is likely to replace Mantoux test. This (C-TB) test is based on ESAT - 6 and CFP – 10 antigens of M-TB and is not affected by prior BCG vaccination status of the child making this test much more specific than the Mantoux test. Use of this test would be similar to the Mantoux test (0.1 ml intradermal injection using the same technique as in Mantoux test and read at 48 - 72 hrs). This test would have a single cut off of >5 mm for both HIV infected and non-infected. The test reagent would come filled in pre filled syringes and the programmatic cost of this test would be INR 30 – 40 per test.

It needs mention that IGRAs (Interferon Gamma Release Assays) also use the same antigens. But in contrast to IGRAs, this test is validated in children below 5 years and is also more robust in patients with low CD4 counts.

Henceforth TST may be used to denoteTB skin test (TST) instead of Tuberculin Skin Test.

4. WRDT – WHO approved rapid diagnostic tests for Tuberculosis

Available WRDT include CBNAAT, LPA and LAMP. A single sample of specimen is sufficient for diagnosis by the more sensitive WRDT. If disease is confirmed by any of these methods, the disease is labelled as microbiologically confirmed TB. CBNAAT is the most easily available WRDT at present.

5. Newer tests

a. Xpert MTB/RIF UltraTMaka CBNAAT Ultra/GeneXpert Ultra: Another WRDT that uses a nested PCR newer generation cartridge which can be used in the same equipment as CBNAAT. The size of the cartridge is larger, therefore a larger amount of sample can be tested. The limit of detection by this test is lower than CBNAAT (16 bacilli per ml of sample vs 131 bacilli per ml) which makes this test more sensitive for detection of TB. This test is especially useful for paucibacillary disease such as for children and HIV co infection. This test is going to replace CBNAAT very soon as they will run on the same CBNAAT machines. Like CBNAAT, Rif resistance can also be detected by this test

b. TruNat: This is another molecular PCR based test for diagnosis of TB and Rifampicin resistance. This test and machine has been developed by an Indian firm MolBioDaignosticsPvt Ltd, Goa. The major advantage of this test is that it is a battery operated machine and can be a point of care test. The disadvantage of this test is that it is not a fully automated test and needs centrifugation of samples like gastric aspirate. It involves a two-step process; DNA extraction and then addition to the DNA chip. This test is still under WHO pre-qualification and is not yet in WRDT.

6. LPA

A test that can be used for genotypic DST. Wider use is recommended because of national strategy of Upfront DST (U-DST). Can be used on smear positive isolates or culture isolates. LPA produces results in 24-48 hours. It is the first and only WHO recommended rapid test for detection of additional resistance in MDR – TB patients as well as X-DR TB.

LPA can detect TB, can detect resistance to R & H (First line LPA) and detect resistance to class FQ (Fluoroquinolones) and class SLI (Second Line Injectables). This helps in decision making on the regime – short VS long MDR, XDR etc.

7. TB – LAMP: Loop mediated Isothermal amplification

This test is recommended as an add on test for detection of pulmonary TB in adults following negative smear microscopy. The sensitivity of this test is higher than that of smear microscopy but lower than that of CBNAAT. This test has several unclear issues for children and for people living with HIV and are not therefore recommended for them. This test cannot replace the rapid molecular tests.

8. TB Culture

Liquid culture has changed the whole scenario of TB diagnostics. It is an important tool for the national strategy for upfront DST. Mycobacterium Growth Indicator Tube (MGIT TM) is now available through RNTCP lab network and other accredited labs. Can be used upfront with specimens such as gastric aspirate, induced sputum and broncho alveolar lavage.

9. UDST

Upfront or universal Drug Sensitivity Testing is the national strategy now. Widespreadpublic-sector deployment of high-sensitivity diagnostic testing and universal DST appropriately linked with treatment could substantially impact MDR-TB in India. Sufficiently wide deployment of Xpert could, moreover, turn an increasing MDR epidemic into a diminishing one. Synergistic effects were observed with assumptions of simultaneously improving MDR-TB treatment outcomes.

10. Major changes in treatment

a. Daily DOTS: The full duration of TB treatment is now daily therapy rather than alternate day therapy.

b. 3 drugs in CP:Isoniazid, Rifampicin and Ethambutol (HRE) instead of HR is to be used in the continuation phase of TB treatment. This is recommended because of the high prevalence of INH mono - resistance and the likely risk of amplifying resistance against Rifampicin.

c. Category II has been withdrawn: Overall success rates of the erstwhile Category II regime was only 60-80% and worse outcomes were seen in patients who were previously treated. There was increased incidence of DR TB in this group.

   Therefore, patients who belong to the following types – microbiologically positive relapse, microbiologically positive failure, microbiologically positive treatment after default, smear negative relapse, recurrence etc. – are all subjected to U DST by WRDT and started on a regime for DR TB or Category I depending on the drug sensitivity status of the patient. This is the current recommendation.

d. Single category: for patients who are –

   i. New microbiologically confirmed drug sensitive pulmonary TB,

   ii. New clinically diagnosed Pulmonary TB,

   iii. New microbiologically confirmed drug sensitive EP TB,

   iv. New clinically diagnosed EPTB,

   v. Drug sensitive previously treated TB (Recurrence, Treatment after loss to follow up, Treatment after failure)

are all treated by 2 HRZE + 4 HRE. Continuation phase is to be extended to 8 months for neuro and spinal TB.

e. Drug dosages have increased:

		Range mg/kg/day	Average mg/kg/day	Maximum mg/kg/day
Rifampicin	R	10-20	15	600
Isoniazide	H	7-15	10	300
Pyrazinamide	Z	30-40	35	2000
Ethambutol	E	15-25	20	1500
Streptomycin	S	15-20	20	1000

f. Usage of FDCs:

WHO and IUATLD recommend usage of FDC formulations as routine practice. They reduce pill burden for children. Fixed drug combinations are safe, simple, reduce the errors of missing one drug thereby reducing the chances of emergence of drug resistant strains. From programmatic viewpoint, they simplify drug supply management, shipping and distribution.

Current FDCs have proper ratios of all drugs and are available in dispersible tablet forms. 3 drug FDC DT has H50, R75, Z150 (10:15:30) for children + non DT Ethambutol 100. 4 drug FDC adult has H 75, R 150, Z 400, E 275.

g. Widened scope of steroid use

The recent guidelines recommend usage of steroids in the following situations, though the quality of evidence is not very robust –

- Miliary TB with alveolo capillary block
- Spinal cord compression with neurological deficit
- Endobronchial obstructive lesion
- Peritoneal TB
- Adrenal TB
- Vasculitis/ Aorto arteritis

h. Pyridoxine supplementation recommended with ATT

Previous guidelines did not recommend routine use of pyridoxine along with ATT except in high risk groups such as in co infection with HIV, malnourished, Diabetes mellitus, renal/liver failure, MDR treatment.

This recommendation has been changed and currently it is recommended to add routine pyridoxine supplementation to all children receiving ATT or IPT. The rationale for this recommendation is that INH doses for ATT and IPT have increased, thereby potentially increasing the risk of dose related adverse effects; high prevalence of malnutrition in children with TB (who are already deficient in pyridoxine); in young children it is difficult to detect and diagnose peripheral neuropathy (which may lead to severe & prolonged morbidity).

Dose:

Prophylaxis - 10mg daily

DR TB – 50 – 100 mg daily depending on the drugs used (Cycloserine may lead to additional risk of peripheral neuropathy)

Evidence of Vitamin B6 deficiency – 50 mg daily

Summary of the new recommendations

- In relevant symptomatology (presumptive TB), one criteria that has been added is "Poor weight gain despite nutritional rehabilitation in children with SAM"
- CBNAAT or other WRDTs should be done if the patient has a TB suggestive Chest X Ray in addition to relevant symptoms rather than on the basis of symptoms only
- New generation TB skin test is going to be introduced
- New WRDTs include CBNAAT Ultra and TruNat will soon be available
- U-DST is national strategy for which LPA

and Liquid culture along with CBNAAT will be the main tools

- Treatment is now daily therapy (DOTS)
- In CP, three drugs HRE is advised even for the new cases
- Category II has been withdrawn – there is now a single category for treatment
- The retreatment cases are to be investigated for drug resistance for definitive therapy
- Scope of steroid use has been widened
- Pyridoxine is recommended for all patients on ATT or IPT

Notification of TB cases is mandatory

DOTS remains the most important component of therapy to maintain the uniformity of regimen, drug dosages and ensuring treatment completion.

Further Readings :

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West Bengal Academy of Pediatrics Activity

STRIDE, organized by WBAP at Hotel Hyatt on 5th January 2019

37 West Bengal Pedicon and 11th National Conference of Computer and Medical Informatics Chapter of the IAP at CII Hall on 8,9 December 2019

Mission Kishore Uday Workshop at Golden Jubilee PEDICON Hall on 23 June 2019

Pediascope, IAP Course of Basic Bronchoscopy at Bhagirathi Neotia, Rajarhat on 25th May 2019

Approach to a Child with Suspected Rheumatological Disorder

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Introduction

Pediatric Rheumatology, though one of the youngest pediatric subspecialities, is evolving at a very fast pace, standing on the pillars of basic immunology, infections and genetics. One of the basic differences of rheumatology from other subspecialities is that instead of involvement of a single organ system, rheumatic diseases are a diverse group of chronic diseases united by the presence of chronic inflammation in multiple systems. Sometimes we, as clinicians, find ourselves not very conversant with elicitation and interpretation of history and clinical examination in a patient with suspected rheumatological disorder, probably because our current curriculums don't give as much importance to rheumatology as a system as is done to other organ systems. Although a thorough knowledge of examination of all organ systems is essential for elicitation and interpretation of rheumatological symptoms and signs, we need to develop our clinical acumen and temperament specific to rheumatology.

Symptoms and signs : Interpreting the Obscure

Rheumatology is often considered "grey area" of medicine as there are only a few useful diagnostic tests, sparse pathognomonic physical tests, controversies even in nomenclature which is ever evolving and therapy that often lacks specificity. Hence a detailed history, thorough physical examination of the whole child with careful observation over time is mandatory. Common mimickers of rheumatologic disorders should also be considered.

Although there is no single classic symptom which would guide a physician to a definite rheumatologic etiology with confidence, the common symptoms include joint pain and swelling, fever, fatigue, and rash.

Chronic or intermittent pain is a primary symptom of many pediatric rheumatic diseases but is often difficult to assess objectively in children. Non verbal clues are more important. Face rating scale is useful (Fig 1). Pain without other physical findings are a frequent reason for referral to pediatric rheumatologists. Arthralgias without physical findings for arthritis suggest infection, malignancy, orthopedic conditions, benign syndromes, or pain syndromes such as fibromyalgia. Arthralgia may also be a presenting symptom of pediatric systemic

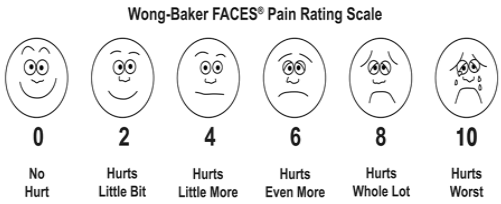


Fig 1. Visual Pain Scale

lupus erythematosus(SLE) and chronic childhood arthritis such as juvenile idiopathic arthritis (JIA). Interestingly, many children with JIA do not complain of joint symptoms at presentation.

Rheumatic diseases may manifest as arthralgias only ,but , objective features of arthritis such as swelling, heat and restriction of movement are stronger predictor to the presence of rheumatic disease. Whenever a swelling is encountered, it is important to assess that swelling is arising from which structure around the joint namely, skin, bursa, tendon, bone tissue or synovial effusion /blood

Fatigue is a nonspecific symptom that may point to the presence of a rheumatic disease but is also common in non-rheumatic causes, such as viral infections, pain syndromes, depression, and malignancy. Fatigue, rather than the specific complaints of muscle weakness, is a common presenting complaint in juvenile dermatomyositis (JDM) . It is also frequently present in SLE,vasculitis, and the chronic childhood arthritis. Overwhelming fatigue with inability to attend school is more suggestive of chronic fatigue syndrome, pediatric fibromyalgia, or other amplified pain syndrome.Table 1 summarises the typical synptoms of some pediatric rheumatological conditions.

A complete physical examination is mandated in any child with suspected rheumatic disease, because many diseases have associated subtle physical findings that will further refine the differential diagnosis. In addition, many rheumatic diseases have multisystem effects, and a stepped assessment should focus on delineating the

Table 1 Common symptoms of some rheumatological conditions

Symptoms	Rheumatic disease(s)	Possible non-rheumatic disorders with similar complinrs
Fever	Systemic JIA, SLE, Vasculitis ARF, MCTD, Sarcoidosis	Malignancies, Infection & postinfections, Inflammatory bowel disease, periodic fever (Autoinflammatory), Kawasaki disease, HSP
Arthralgias	JIA, SLE, JDM, ARF, Scleroderma, Sarcoidosis	Hupothyroidism, trauma, malignancies, growing pain, pain syndrome, overuse syndrome
Weakness	JDM, myositis sccondary to SLE, MCTD, deep localised scleroderma	Muscular dystrophy, Metabolic & other myopathics Hypothyroidism
Chest pain	JIA, SLE (associated with pericarditis or costochondritis	Isolated costochondritis, rib fracture, viral pericarditis, anxiety disorder
Back Pain	ERA, Juvenile AS	Spinal SOL, Vertebral compression fracture, spondylosis, spondylolisthesis, pain syndrome
Fatigue	JIA, SLE, JDM, MCTD, vaculitis	Pain syndrome, Chronic, fatigue syndrome, Depression

JIA - Juvenile idiopathic arthritis, JDM - Juvenile dermatomyositis, SLE - Systemic lupus erythematosus, MCTD - Mixed connective disorder, ERA - enthesitis related, ARF - Acute Rheumatic Fever

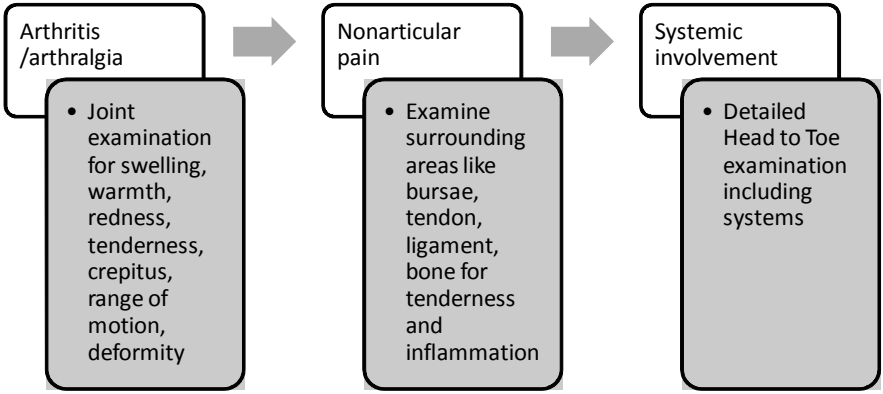


Fig 2. Physical examination depends on the presenting complaints

Table 2 Common and typical physical signs of some rheumatological disorders

Sign	Rheumatic	Comments	Non-rheumatic
Malar rash	SLE, JDM	SLE spares nasolabial fold	Sun burn, Fifth disease, Kawasaki disease
Oral ulcers	SLE, Behcet	Behcet's disease is associated with genital ulcer	HSV infection, PFAFA
Purpuric rash	ANCA positive vasculitis, HSP	HSP rash affects lower limbs and buttocks	Meningococcemia thrombocytopenia, clotting disorder
Gotton papules	JDM	Associated Heliotrope rash, periungual telangiectasias	Psoariasis, eczma

ANCA-Antineutrophilic cytoplasmic antibody, HSV-Herpes simplex virus, PFAFA-periodic fever apthous stomatitis pharyngitis adenitis, HSP-Henoch Schonlein Purpura

extent of organ system involvement (e.g., skin, joints,muscle, hepatic, renal, cardiopulmonary). Table 2 gives a summary of common signs of rheumatologic disorders

Raynaud phenomenon may be a primary benign idiopathic disorder or can be a presenting complaint in the child with scleroderma, lupus, mixed connectivetissue disease (MCTD), or an overlap syndrome. Diffuse lymphadenopathy ispresent in many rheumatic diseases, including SLE, polyarticular JIA, and systemic JIA.

Approach to mono-arthritis :

Acute pain or swelling of a single joint requires immediate evaluation for septic arthritis that can rapidly destroy the joint if left untreated. It is important to distinguish pain from periarticular structures (tendons, bursa), which usually requires only symptomatic treatment, and cases of referred pain (shoulder pain due to peritonitis or heart disease).During history taking trauma must be excluded, and it should provide various clues like Tick Bites (Lyme Disease), sexual risk factors (Giant Cell arthritis), triads like colitis, uveitis, urethritis (Reactive arthritis).

Approach to Poly-arthritis:

Polyarthritis being the commoner complaint

Entity	Aetiology
Infectious arthritis	Batcerial (staphylococcal), Viral (HIV), Lyme's, Mycobacterium, Fungi
Trauma	Fracture, Hemarthrosis
AVN Bone	Trauma, corticosteroid use
Tumours	Primary or metastatic
Systemic disease	Rhuematoid arthritis, reactive arthritis, SLE, psoriatic

in pediatric table practice the evaluation and diagnosis remains of immense importance, the following are some patterns of inflammatory polyarthritis.

So whether the arthritis is symmetric or asymmetric, axial or extra axial that is the pattern of involvement must be delineated first, regarding the etiology, the causes remains vast but majority of them can be snuffed out from a meticulous history taking.

Polyarthritis could be a benign self-limiting illness, the beginning of a serious chronic illness resulting in significant morbidity or a rheumatological emergency requiring urgent intervention. While the differential diagnosis can be very broad, a presumptive diagnosis with regard to the underlying etiology can often

Table 3 Approach to polyarthritis

Pattern	Aetiology
Polyarticular peripheral(usually symmteric)	Rheumatoid arthritis (additive), SLE,Viral arthritis, Psoriatic arthritis
Oligoarticular;axial involvement(usually assymetric, peripheral joints)	Seronegativespondyloarthropathy (Ankylosing spondylitis, Reactive arthritis, Psoriatic arthritis)
Oligoarticular without axial invovlement(usually symmetric joints)	Rheumatic fever(migratory), Lyme disease, enteropathic arthritis,

Table 4 Causes of Polyarthritis

Etiology	Causative agent
Viral	Parvovirus B19, Enteroviruses, Adenoviruses, Mumps, Rubella, Varicella zostervirus, Hepatitis B, Coxsackie virus, Cytomegalovirus, EBV, HIV.
Bacterial	Staphylococcal and streptococcal infections, Neisseria gonorrhae, Hemophilusinfluenzae; Bacterial endocarditis.
Other infections	Tuberculosis, Leptospirosis, Fungal infections, Brucellosis
Parainfectious (reactive)	HIV, Group A streptococcal infections, Salmonella, Shigella, Yersinia,Campylobacter, Mycoplasma,Chlamydia.
Rheumatalogical	Juvenile idiopathic arthritis (JIA), Systemic lupus erythematosus (SLE),Juveniledermatomyositis (JDMS), Behcet syndrome.
Systemic vasculitides	Henoch-Schonleinpurpura (HSP), Kawasaki disease (KD), Polyarteritisnodosa (PAN), Wegener's granulomatosis
Spondyloarthropathies	Juvenile ankylosing spondylitis (JAS), Psoriatic arthritis Enteropathic arthritis.
Miscellaneous	Sarcoidosis, Drug/serum sickness reactions

be made by careful attention to the clinical history and physical examination

Examination and Investigation :

It is said that 80% of the rheumatological diagnosis comes from clinical history, 15% from examination and only 5% from investigation.So apart from the history taking, examining the patient is also important just to differentiate between a chronic disease in early phase or just a pain syndrome requiring symptomatic treatment. Few areas need to be given special emphasis during clinical examination like checking the vitals including the blood pressure with an appropriate size cuff, detailed anthropometry which will give us an idea about the duration of the disease, and thorogh examination of hair(alopecia), skin(helitrophic rash,Gottron's papule, discolouration, calcinosis),nail (pitting, periungual telangiectasia),nose and mouth(oral ulcers, red tongue,cracked lips),genitals(ulceration)back(spinous process, paraspinal and surrounding muscle,sacroilliac joint)

Detailed systemic examination would include examination of cardiovascular and respiratory systems (pericardial and pleural rub, decreased breath sound due to pulmonary fibrosis) and examination of abdomen for ischemic pain, hepatosplenomegaly and ascities. It is the constellation of physical signs rather than a single finding that provide clues for diagnosis on the basis of preset disease specific diagnostic criteria.

There are two system available in examining a patient of presumed rheumatic disorders i.e.

pGALS(paediatric Gait, Arms, Legs, Spine) as screening test and

pREMS (paediatric regional examination of musculoskeletal system) as detailed examination.

pGALSis a simple screening approach to musculoskeletal(MSK) examination which can be done in a very short time (<2 minutes) in school-aged children and may be successfully performed in younger ambulant children as well. It is a simple and quick test(average 2 mins) having an excellent sensitivity to detect abnormality (>97%) and acceptability by children and their parents.

It is different from adult GALS screen though the sequence of pGALS is essentially the same as adult GALS with additional maneuvers to screen the foot and ankle (walk on heels and then on tiptoes), wrists (palms together and then hands back to back) and temporomandibular joints(open mouth and insert three of the child's own fingers),and with amendments at screening the elbow (reach up and touch the sky) and neck (look at the ceiling). These additional maneuvers were included because when adultGALS was originally tested in school-aged children it missed significant abnormalities at these sites.

When to Perform pGALS

(a) Child with muscle, joint or bone **pain**

(b) Unwell child with pyrexia

(c) Child with **limp**

(d) Delay or regression of **major milestones**

(e) **'Clumsy'** child in the absence of neurological disease

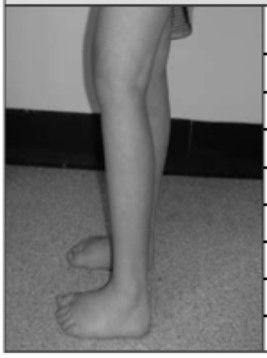
(f) Child with **chronic disease** and known association with MSK presentations.

Foster HE, Kay LJ, Friswell M, et al. Musculoskeletal screening examination (PGALS) for school-age children based on the adult GALSscreen. Arthritis Rheum 2006: 15,55;981

pGALS – A musculosketal screening assessment for school-aged children	
Screening Questions	• "Do you have any pain or stiffness in your joints, muscles or back ?" • "Do you have any difficulty getting yourself dressed without any help?" • "Do you have any difficulty going up and down stairs ?"
GAIT	• Observe the child walking • "Walk on your tip-toes/walk on your heels"
ARMS	• "Put your hands out in front of you" • "Turn your hands over and make a fist" • "Pinch your index finger and thumb together" • "Touch the tips of your fingers with your thumb" • "Squeeze the matacarpophalangeal joints" • "Put your hands together/put your hands back to back" • "Reach up and touch the sky" • "Look at the ceiling" • "Put your hands behind your neck"
LEGS	• "Feel for effusion at the knee" • ""Bend and then straighten your knee" (check active movement of knees and feel for crepitus) • Passive flexion (90 degrees) with internal rotation of hip
SPINE	• "Open your mouth and put 3 of your (child's own) fingers in your mouth" • Lateral flexion of cervical spine – "Try and touch your shoulder with your ear" • Observe the spine from behind • "Can you bend and touch your toes?" Observe curve of the spine from side and behind.

Documentation of the pGALS screen

Documentation of the pGALS screening assessment is important and a simple pro forma is proposed with the following example – a child with a swollen left knee with limited flexion of the knee and antalgic gait.



pGALS screening questions		
Any pain?	*Left knee*	
Problems with dressing?	*No difficulty*	
Problems with walking?	*Some difficulty on walking*	
	Appearance	Movement
Gait		✗
Arms	✓	✓
Legs	✗	✗
Spine	✓	✓

Helen E Foster and Sharmila Jandial, Pediatric Rheumatology201311:44,https://doi.org/10.1186/1546-0096-11-44

Some practical tips for pGALS include

- Get the child to copy you doing the manoeuvres
- Look for verbal and non-verbal clues of discomfort

  (e.g. facial expression, withdrawal)
- Do the full screen
- Look for asymmetry (e.g. muscle bulk, joint swelling, ROM)
- Consider clinical patterns and association of leg-length discrepancy and scoliosis

Rule of "6" for variation in knee alignment:

If the child is under 6 years old and there is less than 6 cm between the knees (if has bow legs) or 6 cm between the ankles (if has knock knees) then the problem is likely to be self-limiting

Those with abnormal findings in pGALS must undergo.

pREMS(pediatric Regional Examination of Musculoskeletal System) which is a detailed local examination based on principles of " look, feel and move".

Points to Remember

- In a child presenting with symptoms suggestive of musculoskeletal system, first do pGALS.
- This gives clues to sites requiring further detailed examination.
- Identify rheumatological from non rheumatological illness.
- Identify inflammatory from non inflammatory aetiology.
- Identify articular from non articular causes.
- Identify pattern of joint affection
- Detail systemic examination is essential.
- Constellation of signs and symptoms and disease evolution over time helps clinch diagnosis and order appropriate investigations.

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Neonatal Pseudohypoparathyroidism

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Abstract

The case of a neonate is presented who had early onset hypocalcemia, hyperphosphatemia, and raised parathyroid hormone. The infant did not have any stigmata of pseudohypoparathyroidism. The hypocalcemia was initially resistant to calcium therapy, but responded to vitamin D analog therapy. The diagnosis of 'neonatal pseudohypoparathyroidism' was entertained; the infant remained stable with normal serum biochemistry during 15days of follow-up .E-mail: dr_viveksharma@yahoo.com

Pseudohypoparathyroidism (PHP) is a heterogeneous group of disorders characterized by hypocalcemia, hyperphosphatemia, increased serum concentration of parathyroid hormone (PTH), and insensitivity to the biological activity of PTH[1]. A Medline search revealed very few case reports of PHP presenting as early onset neonatal hypocalcemia. We report a case of pseudohypoparathyroidism that presented in early neonatal period with hypocalcemia.

Case Report

A preterm male neonate born by vaginal route was found to have hypocalcemia on routine screening for prematurity. The baby, born to a 32-years-old primigravida at 31week 4days gestation, cried soon after birth and was transferred to neonatal intensive care unit for further management. The parents were healthy with normal stature with history of consanguinity. mother conceived by IVF had placentae previae and hypothroidism.

The baby weighed 1.67 kg, with head circumference of 33 cm. Anterior frontannele was at level and there was no dysmorphic facies or gross congenital malformation. Rest of the systemic examination revealed no abnormality.

Provisional diagnosis of prematurity and low birth weight was made and investigations revealed blood glucose 87 mg/dL and serum calcium 5.8 mg/dL. Blood urea was 9 mg/ dL and septic screen analysis was negative. Ultrasound skull and abdomen did not reveal any abnormality. In view of hypocalcaemia i.v. calcium gluconate was started at 8ml/kg/day. Despite that calcium remained low at 48 hours of calcium infusion. Persistence of

hypocalcemiae led to further investigations which revealed serum calcium – 6.5 mg/dL, serum phosphorus – 7.9 mg/dL(normal: 3.8-6.5 mg/ dL) and alkaline phosphatase of 162 IU/l (normal 150-400 IU/l). Repeat blood urea was 32 mg/dl and serum creatinine, 0.2 mg/ dL. The serum parathormone levels by radioimmunoasssay was 113pg/mL (normal: 10-70 pg/mL), with simultaneous serum calcium of 5.9 mg/dL. Serum calcium, phosphorus and alkaline phosphatase of mother were normal. In view of persistent hypocalemia, hyperphosphatemia and high serum parathormone levels, diagnosis of PHP was made, and the baby was treated with calcium supplementation and vitamin D analog, calcitriol in a dose of 0.25 µg/day[2,4]. Serum calcium and phosphorus profile improved after starting calcitriol. On follow up at 15 days sr. calcium was 7.3 and sr. phosphorus was 7.1. the child was asymptomatic on calcitriol and calcium supplementation. Further plan is to continue same treatment till calcium and phosphorus normalizes. After that treatment can be ceased temporarily to see if the condition is transient or permanent PHP as transient PHP gets resolved within 6 monthsof age[3].

Discussion

In 1942, Fuller Albright first introduced the term pseudohypoparathyroidism to describe patients who presented with PTH-resistant hypocalcemia and hyperphosphatemia. In PHP, the parathyroid glands are normal or hyperplastic histologically, and neither endogenous nor administered PTH raises the serum levels of calcium or lowers the level of phosphorus[1].

Pseudohypoparathyroidism is divided into 2 main types. Type I is characterized by low or absent renal cyclic adenosine monophosphate (CAMP) production in response to parathormone (PTH). Type II responds to PTH with normal increase in urinary CAMP but shows absent or subnormal phosphaturic response2. Type I is further subdivided into 2 subtypes, A and B. In sub type A, the affected patients have a genetic defect of the a subunit of the stimulatory guanine nucleotide binding protein (GSa), with most of them having distinctive morphological abnormalities collectively called “Albrights herediatry osteodystrophy”.1 In this type, hypocalcemia rarely develops before 3 years.3 Subtype I B patients have normal levels of G protein activity with defect in PTH receptor expression or a defect in catalytic subunit of adenyl cyclase.

In our case, the baby presented in the early neonatal period with hypocalcemia detected incidentally during screening which was resistant to i.v. calcium gluconate. Septicemia and renal failure were ruled out. Elevated levels of serum parathormone levels further ruled out hypoparathyroidism. Case reports of transient PHP that presented as late onset hypocalcemia are available in the literature but to the best of our knowledge there are only few case reports in literature of pseudohypoparathyrisdism presenting as early onset hypocalcaemia[4]. At 15 days of life baby was asymptomatic on oral calcium supplementation and calcitriol and had improving parameters.

All patients with hypocalcemia whether symptomatic or asymptomatic should be initially treated with intravenous or oral calcium. Administration of oral calcium and 1 alphahydroxylated vitamin D metabolites, such as calcitriol, remains the mainstay of treatment and should be initiated in every patient with a diagnosis of PHP. The goals of

therapy are to maintain serum total and ionized calcium levels within the reference range to avoid hypercalciuria and to suppress PTH levels to normal. This is important because elevated PTH levels in patients with PHP could cause increased bone remodeling and can lead to hyperparathyroid bone disease.

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