Karnataka Paediatric Journal Vol. 29, No. 2 April - June 2014

Journal of the IAP Karnataka State Branch

CONTENTS

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RICKETTSIAL FEVER- AN INCIDENCE STUDY IN A TERTIARY CARE CENTRE IN SOUTH INDIA

Dr. NirmalaVineetha Pinto , Dr. Sudha Rudrappa, Dr. Suresh R.

ABSTRACT:

Rickettsial Fever is a zoonotic febrile illness spread by bite of ticks and mites.

This study was done to measure the incidence of rickettsial fever in Pediatric ward a tertiary care hospital in Mysore

This study included all children between the age group 1year to 14 years who were admitted in Cheluvamba Hospital Mysore from 1st July 2012 to 31st June 2013 .The Case records of all children with a clinical diagnosis of rickettsial fever and who tested positive for Weil Felix reaction were reviewed. Weil Felix test was done by the tube agglutination method .A titre of 1/ 160 and above was considered significant. RGA Scoring system for spotted fever group of Rickettsial fever was used (Score above 9 was taken as positive).Statistical Analysis used was SPSS version 17 for windows and data analysis was by descriptive analysis and chi square test. In our study a total of 13 children (4.56% of viral exanthematous fever) were found to have rickettsial fever. Age ranged from 1 y to 14 years with maximum incidence below 5 years (46.2%) and male predominance (M:F = 1.6:1). Major presenting symptoms were fever (100 % with mean of 8.76 days) and rash (100%), followed by oedema (38.5%), hepatosplenomegaly (21%).

CONCLUSIONS :

Rickettsial fever should to be considered in children with presenting complaints of fever with rash. A positive Weil Felix reaction in the presence of clinical suspicion and the RGA scoring above 9 can be regarded as diagnostic for rickettsial fever where facilities for confirmatory tests are not available. Early diagnosis and treatment is associated with reduced morbidity and mortality.

KEY WORDS :

Rickettsial Fever, RGA scoring system, Weil Felix reaction.

INTRODUCTION:

Rickettsial Fever is an acute febrile zoonotic illness transmitted by bite of mites and ticks with varied clinical manifestations presenting as fever with rash or accompanied with gastrointestinal, respiratory orcentral nervous system involvement. The main agent causing rickettsial infection in our country is Orientiatsutsugamushi,which is the etiological agent for scrub typhus[1].Often labeled as viral 'Exanthematous fever''[2]. Rickettsial infection can lead onto serious end organ damage which includes pneumonitis, ARDS, acute renal failure, myocarditis, disseminated intravascular coagulation (DIC)[3] and septic shock.[4] The

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presence of Rickettsial fever and its re-emergence have been established in various parts of India- Jammu and Kashmir, Himachal Pradesh, Uttaranchal, Rajasthan, Assam, West Bengal, Maharashtra, Kerala, Tamil Nadu and Karnataka.

Diagnosis of the disease is often challenging due to non-availability of confirmatory laboratory tests and cost factor.

Diagnosis and confirmation of this disease is based on Serological tests like Micro Immunofluorescence, Latex agglutination, Indirecthemagglutination, ELISA and Immunoperoxidase[3] .Gold standard test for diagnosis isimmunofluorescence assay.

The Weil-Felix (WF) test is based on the detection of antibodies to various Proteus species which contain antigens with cross-reacting epitopes to antigens from members of the genus Rickettsia. Whole cells of Proteus vulgaris OX-2 react strongly with sera from persons infected with SFG rickettsiae,and whole cells of P. vulgaris

OX-19 reacts with sera from persons infected with typhus group rickettsiae.

The OX-K strain of Proteus mirabilis was demonstrated to agglutinate with sera from scrub typhus patients5. By the WF test, agglutinating antibodies are detectable after 5 to 10 days following the onset of symptoms, with the antibodies being mainly of the immunoglobulin M (IgM) type. The poor sensitivity of the WF test is now well demonstrated but a good correlation between the results of the WF test and detection of IgM antibodies by an immunofluorescence assay (IFA) is often observed.

The RGA scale was devised by Rathi GoodmanAghai et al. includes clinical and laboratory findings in scrub typus[6].Total score is 35 (25 clinical and 10 laboratory).

TABLE 1 shows the RGA scoring system for spotted fever

Clinical features	Score	Laboratory	Score
Rural	1	Hemoglobin <9 g/dl	1
Pets	1	Platelets< 1,50,000	1
Tick Exposure	2	CRP> 50 mg/dl	2
Tick Bite	3	Serum Albumin < 3g/dl	1
Conjunctival congestion (Non Exudative)	2	Urine albumin > 2+	1
Maculopapular Rash	1	SGPT> 100 (U/L)	2
Purpura	2	Na <130 (mg/L)	2
Palpable purpura/ ecchymosis/necrotic rash	3		
Rash appearing 48-96h after fever	2		
Pedal edema	2		
Rash on palm and soles	3		
Hepatomegaly	2		
Lymphadenopathy	1		

criteria. Age ranged from 1 year to 14 years with maximum incidence below 5 years age group (46.2%) and male predominance(M:F=1.6:1).

Major presenting symptoms were fever (100 % with mean of 8.76 days) and rash (100%), followed by oedema (38.5%), hepatosplenomegaly (21%).

TABLE- 2 Observations of the study

S. N	AGE	SEX	FEVER (DAYS)	RASH/EDEMA	HSM	WEIL FELIX		RGA SCORE	DOXY DURATION
						OX-2	OX-19		
1	7yrs	M	7	RASH-1DAY	NIL	1:320	1:320	11	4DAYS IMPROVED
2	1yr 2mn	M	10	BOTH-3DAYS	NIL	1:180	1:320	14	3DAYS IMPROVED
3	11yrs	F	8	RASH-8DAYS	NIL	1:180	1:320	13	5DAYS IMPROVED
4	4yrs	M	7	BOTH-7DAYS	NIL	1:180	1:320	15	5DAYS IMPROVED
5	1yr 6mn	F	8	BOTH-3DAYS	NIL	1:160	1:160	15	6DAYS IMPROVED
6	12yrs	M	1	RASH-3DAYS	NIL	1:160	1:160	14	3DAYS IMPROVED
7	2yr	M	15	RASH-3DAYS	NIL	1:160	1:160	14	4DAYS IMPROVED
8	1yr	M	15	RASH-3DAYS	NIL	1:160	1:160	11	5DAYS IMPROVED
9	3yr	F	7	RASH-1DAY	NIL	1:160	1:160	11	3DAYS IMPROVED
10	15yrs	F	9	RASH-2DAYS	Hpm-2cm	1:160	1:160	21	5DAYS IMPROVED
11	11yrs	M	15	RASH-2DAYS	Hpm-2cm	1:160	1:160	14	5DAYS IMPROVED
12	6yr	M	8	BOTH-1DAY	NIL	1:160	1:160	15	3DAYS IMPROVED
13	8yr	F	4	RASH-1DAY	NIL	1:180	1:640	11	5DAYS IMPROVED

On Weil Felix test, 8 children showed titres = 1:160 for OX2 antigen and 8children for OX-19 antigen = 1:160, while 4children showed titre =1:320 for both the antigens at presentation 1child showed a titre of 1:640. RGA scores had a median of 14.

DISCUSSION:

This study documents that rickettsial fever is prevalent in Karnataka and needs to be considered as an essential differential diagnosis in children presenting with exanthematous fever. In our study 13 children met our inclusion criteria and had a positive Weil Felix test and RGA scores were above 9.Incidence being 4.29 % with a male preponderance and maximum in age group less than five years. Weil Felix test even with low sensitivity can be used in a set up where confirmatory tests are not available,however it should be correlated with clinical features and the RGA scoring system can be applied.

The clinical features indicating strong suspicion for diagnosis includes rashes involving palm and soles, edema of hands and soles, hepatosplenomegaly and arthralgia.

The importance of rapid diagnosis is the prompt response to antimicrobial therapy and reduced mortality in children with early treatment. The treatment given in our set up was oral doxycycline 4 mg/kg/ day7till 3 days after being afebrile, to which all the children responded and there was no morbidity.

Delay in diagnosis and treatment has been associated with death and grave neurological sequelae.

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KARNATAKA IAP PRESIDENT ELECTION 2016

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SOCIOECONOMIC STATUS AMONG SCHOLASTIC BACKWARD CHILDREN

Dr Nagarathna, Dr Anitha S

ABSTRACT

The present study compares influence of socioeconomic status on scholastic backwardness of children from rural and urban background.

The research collected data from students belonging to both rural and urban schools by administering a questionnaire to 223 students. Students were selected based on a systematic random sampling method and belonged to the age group 6 to 12 years. Questionnaire was provided to teachers included demographic details and questions related to their perceived level of scholastic capability. The data collected were subjected to statistical analysis using SPSS software.

The study showed that there is a significant association between SES and academic achievement among urban and rural children ($p<0.001$) while the boys displayed higher prevalence of scholastic backwardness compared to girls.

CONCLUSION:

A significant relationship between socioeconomic class and scholastic backwardness among urban and rural was observed.

KEY WORDS :

Scholastic backwardness, Academic achievement, Socio economic status, urban, rural Children, India.

INTRODUCTION

In today's competitive world, academic underachievement of children is a significant concern among parents and teachers. Scholastic backwardness in terms of poor academic achievement or repeated failure in grades is being increasingly recognized as one of the important problems in children . Recent studies conducted in the past recorded the prevalence of scholastic backwardness in Indian school children ranges between 20 -60% (1-2) which alarms the society as a whole. In addition, the scholastic difficulties have been associated with school absence , poor concentration, poor school functioning , adverse family conditions , father's occupational status and large family size (3). Among different factors, socio-economic status (SES) was found to be associated more strongly as shown consistently in previous studies. A disadvantaged child because of social or cultural factors including social class or poverty was likely to possess less or poor knowledge, attitude and skills required for the school system which in turn makes adjustment difficult and impedes learning .

Several studies conducted among school students in the past have showed a positive relationship between SES and academic achievement .High SES was found to associate with high achievement according to Sharma . This was due to the assumption that the high SES families are

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presumed to be highly educated than their counterparts . It is also ben observed that students belonging to upper SES significantly scored better academic achievement than lower status. In addition to the SES background, researchers have also conducted with specific reference to sub-cultural groups showing that the economically disadvantaged showed poor achievement than the advantaged children . In general, these studies imply that apart from SES of the children, families living in rural and urban communities, on average, are poorer than their suburban counterparts. Thus, it is important to account these differences when comparing scholastic achievement of children.

Although the previous studies documented the role of socio-economic status and cultural impact on academic achievement, but all these studies have been conducted in the year of early eighties.(4) There was a significant change from the past in terms of academic skills such as reading, writing and numeric due to the several changes such as urbanization, role of technology especially internet revolution and globalization. Hence, these findings perhaps may not be applicable to today's scenario. Furthermore, a recent study by Kamble and Takpere assessed the scholastic backwardness among third grade students but the study did not account the SES. Hence, with this background the present study was conducted to assess the impact of location on (rural or urban area) on scholastic achievement. In addition, the study also determines the impact of both a family's socioeconomic status on child's academic achievement.

MATERIALS AND METHODS

A cross-sectional study with descriptive research design was conducted at five rural and five urban schools of Mangalore, India to ensure that both socio-economic statuses were included in the present study (5).

2nd standard to 7th standard school children from each area (urban and rural were taken up for studies. Questionnaires were administered to each student which contained demographic questions, socioeconomic status and questions related to students' perceived scholastic level. The scholastic backwardness was assessed based on Rutters Proforma A, and grades obtained in the previous two examinations. SES of scholastic backward children assessed by modified Kuppuswamy scale.

RESULTS

In total 106 from rural and 117 scholastic backward children from urban area were participated in the study. Table 1 presents the demographic characteristics of the children participated in the study.

Table 1 : Descriptive statistics for demographic profile of the children

	Location	Total	Chi-square value
	Rural (n=106)	Urban (n=117)	
Sex	0.095	0.757	
Girls	42(39.6%)	44(37.6%)	86(38.6%)
Boys	64(60.4%)	73(62.4%)	137(61.4%)
Total	106(100.0%)	117(100.0%)	223(100.0%)
Age group	3.694	0.158	
<= 9	50(47.2%)	42(35.9%)	92(41.3%)
10 - 11	30(28.3%)	46(39.3%)	76(34.1%)
>=12	26(24.5%)	29(24.8%)	55(24.74%)
Total	106(100.0%)	117(100.0%)	223(100.0%)
Medium of instruction	100.339	0.001	
English	0 (0.0%)	74 (63.2%)	74 (33.2%)
Kannada	106 (100.0%)	43 (36.8%)	149 (66.8%)
Total	106 (100.0%)	117(100.0%)	223 (100.0%)

The findings revealed that there is no significant difference in the gender and age group between urban and rural. However, majority of the students belong to English medium in urban while none in rural and these differences found to be significant ($p<0.001$). Thus, medium of instruction varies according to the location.

Table 2: Prevalence of scholastic backwardness children based on Socio economic status and Location between urban and Rural

Socioeconomic status	Location	Total	
	Rural	Urban	
Upper	0	1	1
	0.0%	0.9%	0.4%
Upper Middle	2	32	34
	1.9%	27.4%	15.2%
Lower middle	28	56	84
	26.4%	47.9%	37.7%
Upper lower	50	28	78
	47.2%	23.9%	35.0%
Lower	26	0	26
	24.5%	0.0%	11.7%
Total	106	117	223
	100.0%	100.0%	100.0%

The Table 2 presents the prevalence of scholastic backwardness among urban and rural within different socio-economic status. It is clearly observed that prevalence of scholastic backwardness was higher in rural, particularly in lower (24.5%) and upper lower (47.2%) socio economic status group while this was 23.9% in upper lower and none in lower SES in urban location. Further, interestingly the findings also revealed that 0.9% (n=1) children had scholastic backwardness in urban while this was none in rural, especially in upper income group. Thus, the prevalence was higher within in lower socioeconomic strata presented in rural area than in urban area ($p<0.001$), which further implies that there is a significant association between the socioeconomic status and location.

DISCUSSION

A number of factors contribute to scholastic backwardness of student. These factors can be external which are environmental or psychological problems faced by students. Factors affecting scholastic achievement of students include below average intelligence, physical illnesses,attention deficit hyperactivity disorder, learning disorders, family support and school factors (6). In this study, a total sample size of 223 students of age 6 to 12 years were investigated of which 106 students were from rural area and 117 students were from urban area. The scholastic ability of students who belonged to different socioeconomic class were analysed based on data collected through questionnaire. The different socio economic classes taken into consideration were upper class, upper middle class, lower middle class, upper lower middle class and lower class. The present study has analysed the relationship between socioeconomic status and students in rural and urban background.

The frequency of scholastic backwardness among rural students was greater than urban students. Mishra and in his studies observed that there is significant correlation between socioeconomic status and academic achievement among students. The present study is line with the previous study where a significant positive association between SES and academic achievement among urban and rural students were observed. Contradicting to these results, Narang (5) did not find any relationship between SES and academic achievement among both

rural and urban school students. Further, a recent research suggests that there is a strong correlation between social backgrounds; geographic location and numeracy ability of students indicating that socioeconomic background affect mathematical skills of students (7). However, this difference could be because Mishra did not differentiate between rural and urban school students. Further, no significant relationship between socioeconomic class and scholastic backwardness was observed between urban student population and rural student population. The result obtained in the present study is similar to Kasthuria , Girijia who have showed a significant impact of SES on academic achievement among urban and rural students on an average, are poorer than their suburban counterparts. In this study, scholastic backwardness was still persistent in upper socioeconomic status group but high percentage of scholastic backwardness children in urban area belong to lower middle and upper middle group.

In addition, scholastic backwardness was found to be less prevalent among girls in the sample population compared to boys. This result is different from a study that showed that boys exhibited higher perceived self competency compared to girls among a Caucasian population . This difference could be due to difference in ethnicity and cultural differences. The present study took into consideration a sample population that received two different medium of instructions - Kannada and English, of which most students learnt lessons in Kannada medium. All English medium students belonged to the urban population. This discrepancy may also be a reason for difference in prevalence noted as compared to previous studies.

CONCLUSION

The study concludes that socioeconomic status does influence scholastic backwardness & the impact is more on rural children . However, there are certain limitations to this study: The study employed a small sample size and focused on a small region which questions the generalizability of the results. Time constraint was a primary limitation for this study and hence failed to employ different methods of measuring scholastic levels of students. Future research should therefore employ a larger sample size with diverse population.

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DYKE-DAVIDOFF-MASSON SYNDROME - A CURIOUS CLINICAL DIAGNOSIS.

Dr. Mahesh Maralihalli. Dr. Mahantesh Matti

ABSTRACT:

A 14 year old male child presented with status epilepticus, upper motor neuron type of hemiparesis witn ipsilateral facial weakness, microcephaly. Child had global developmental delay with motor milestones more delayed than mental milestones. CT of the brain revealed features suggestive of Dyke-Davidoff-Masson syndrome.

KEY WORDS:

Dyke-Davidoff-Masson syndrome, cerebral hemiatrophy, hemiparesis

INTRODUCTION:

Dyke-Davidoff-Massonsyndrome (DDMS) refers to cerebral hemiatrophy, which is a consequence of an injury to the brain during fetal period or early childhood (1). Its typical radiological features are skull vault thickening (compensatory), elevation of the petrous ridge, hyper pneumatization of the frontal sinus (also ethmoidal and mastoid air-cells), ipsilateral falcine displacement. The cause of hemiatrophy may be capillary malformations(2), hemispheric infarction etc.

These children present with seizures, poor school performance, facial asymmetry, spastic hemiplegia. There also can be affection of sensory system and psychiatric symptoms(3). The typical clinical features are seen in adolescents and adults (4). However, it can also be seen in children (5). We present a 14-year-old male child with typical clinical and imaging features of DDMS.

CASE REPORT :

14 year old male, only child born to a non-consanguineously married couple, presented with uncontrolled seizures since 2 hours to emergency. Child was treated like status epilepticus, seizures controlled after loading with Inj Phenytoin. Past history revealed child was born by full term normal vaginal, hospital delivery. Baby cried immediately after birth, birth weight was 3 kg. Post natal period was uneventful. He was discharged after 3 days. Child had global developmental delay, motor delay was more affected than mental mile stones. Presently child is being able to walk and run but with weakness on right half of the body. School performance is poor with mild mental retardation. Examination revealed stable vitals, microcephaly, right sided upper motor neuron type of hemiparesis with ipsilateral facial weakness. Other cranial nerves examination was normal. There were no neurocutaneous markers. Vision and hearing was normal. Other systems were normal. Plain 128 Slice MDCT Scan of the Brain was performed with 5 mm thickness

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axial slices which revealed old infarct with gliosis in left fronto-parietal regions with resultant exvacuo dilatation of ipsilateral lateral ventricle, mild calvarial thickening of right parietal bone, hyper-pneumatization of left frontal sinus, mild hypoplasia of left petrous apex. These features are consistent with Dyke Davidoff mason syndrome. He was discharged with oral valproic acid and was given physiotherapy. Child is seizure free till now, 2 months after discharge.

DISCUSSION:

It was initially described by C.G Dyke, L.M Davidoff and C.B Masson in 1933 (6). This syndrome is based on classical features on neuroimaging and typical clinical features. Its radiological features are compensatory skull vault thickening, hyperpneumatisation of frontal sinus,petrous ridge elevation andipsilateralfalcine displacement (7). Clininal features are spastic hemiparesis, facial weakness, mental retardation and seizures. There can be problems of speech, learning disabilities etc.

The radiological findings occur as a result of an insult to the growing brain. As brain does not grow properly, there occurs wideneing of diploic spaces, sinuses enlarge and petrous ridge elevation (8). Infections, ischaemia, trauma, tumor, hemorrhage, vascular anomalies that can result in these manifestations.

In our case, child presented to us at 14 years of age with symptoms that were noticed since 1 year of age. Child was not properly evaluated and diagnosed till now. Probably child might have suffered an insult to brain during fetal stage with radiological evidence of infarct in brain. Child has spastic hemiparesis, mental retardation, facial asymmetry and seizures.

Differential diagnosis considered were hemiplegic cerebral palsy, basal ganglia germinoma and Rasmussen encephalitis. Detail history, physical examination and classical neuro-radiological findings confirm diagnosis (7).

These patients need multidisciplinary treatment. Aggressive control of seizures is must to ensure good outcome. These children need continued long term follow ups in speech therapy and physiotherapy. Multiple anticonvulsants may have to be used sometimes to control seizures. Surgical intervention of choice in intractable seizures is hemispherectomy. Absence of intractable seizures and onset of hemiparesis after 2 years are good prognostic factors (9).

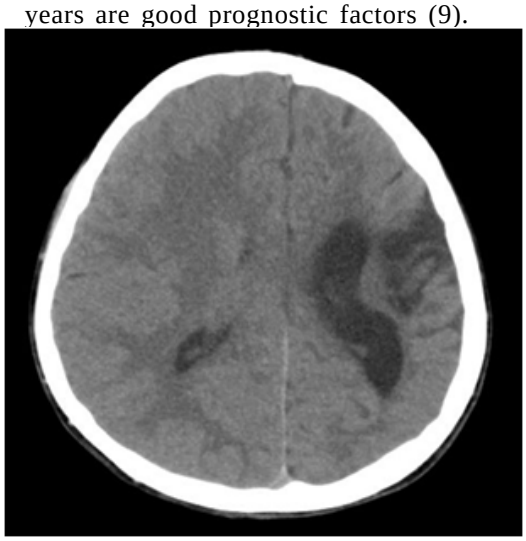

Figure 1 : plain CT showing Old infarct with gliosis in left fronto-parietal regions with resultant exvacuo dilatation of ipsilateral lateral ventricle. . Hyper-pneumatization of left frontal sinus.Mild hypoplasia of left petrous apex.

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FORM IV

1. Place of Publication	:	Mangalore
2. Periodicity of its Publication	:	Quartterly
3. Printerís Name	:	Dr. B. Sanjeev Rai
Nationality	:	Indian
Address	:	Medicare Centre, Mangalore - 575 003
4. Publisherís Name	:	As above
Nationality		
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5. Editorís Name	:	As above
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A STUDY OF CLINICAL, BACTERIOLOGICAL PROFILE AND THERAPEUTIC OPTIONS IN EMPYEMA THORACIS IN CHILDREN

Dr Srinivasa, Dr Sreeja Kottapally , Dr Radhana.S , Dr. Shreedhar Murthy.

ABSTRACT :

Childhood empyema is an important complication of bacterial pneumonia and has significant morbidity and mortality .There are no standard guidelines for management of empyema thoracis.The management remains controversial in terms of duration of antibiotic therapy and the indications for and timing of surgery. This study emphasizes that in tertiary care centres ,VATS can be preferred over medical therapy in appropriate cases for the management of empyema thoracis.

INTRODUCTION :

Childhood empyema is an important complication of bacterial pneumonia and has significant morbidity and mortality. The aim of treatment is to ensure rapid recovery with a normal long term pulmonary outcome. Medical therapy includes use of antibiotics and chest tube drainage .Newer modalities of treatment includes early primary surgical intervention called VATS(Video Assisted Thoracoscopic Surgery) which is associated with lower mortality, lesser ICD days, and reduced incidences of reintervention. There are no standard guidelines for management of empyema thoracis.The management remains controversial in terms of duration of antibiotic therapy and the indications for and timing of surgery. This study emphasizes that in tertiary care centres ,VATS can be preferred over medical therapy in appropriate cases for the management of empyema thoracis.

METHODS

This was a descriptive clinical evaluation study conducted over a period of 20 months.All children between 1 month to 15 years who were admitted with empyema were included in the study.The diagnostic criteria was presence of pus on thoracocentesis and if pleural fluid is nonpurulent ,a positive gram stain / a positive culture.Patients with transudative causes of pleural effusions, postsurgical empyema ,posttraumatic empyema and immunodeficiency were excluded.

A detailed history with emphasis on duration of symptoms was taken and examination done.Later chest X ray and USG (if required) were done.In all clinically suspected cases a diagnostic thoracocentesis was performed and the obtained fluid was sent for analysis.Routine investigations were done in all cases.

Patients with severe distress underwent immediate ICD and those who were not, were managed with a stepwise approach.Those diagnosed to have non loculated empyema on USG

underwent ICD.ICD tube sizes were selected based on child's age and fluid viscosity .Initially they were started on IV amoxicillin-clavulanic acid and amikacin.If Staphylococcal pneumonia was suspected,vancomycin infusion was given.Appropriate antibiotics were added according to culture sensitivity reports.

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Chest X ray was done after ICD insertion to confirm the position of the tube.Daily bedside examination was done to check for vitals,air entry & ICD tube patency.Whenever blockage was suspected,intercostal tube was readjusted.Chest physiotherapy was started at the earliest.All of them were encouraged to blow balloons /use incentive spirometry for good lung expansion.ICD was removed when the drainage was less than 50ml/day with clinical & radiological improvement.Children with persistent symptoms,incomplete lung expansion on ICD & antibiotics,multiple loculations & thick pleural peel on USG on admission/ developed any time during their hospital stay were subjected to surgical line of management.Thoracoscopic breaking of loculi was done in patients with minimal loculations & no thick peel,whereas thoracotomy & decortications was done for those with multiple loculations & thick peel.Appropriate antibiotics were given for a minimum of 4-6 weeks.Patients were discharged after confirming good lung expansion & absence of fever.

All the patients were followed up after 1 & 3 months,and were assessed for lung expansion & chest wall deformities.

RESULTS

During the study period 40 patients were evaluated, with 17 (42.5%) in the age group of 1-4 yrs. Male to female ratio was 1.5:1. 31 patients(77.5%) belonged to the lower socioeconomic group. Prevalence was more during October to March i.e winter to spring(57.5%). Fever & cough were noted in all patients.23 patients presented with right sided & 17 with left sided empyema . Hurried breathing was seen in 72.5% cases. Patients presented to the hospital with a mean duration of 11 days since the onset of symptoms. Pneumonia was predisposing factor in 38(95%) of cases. Pyogenic liver abscess rupturing into the pleural cavity was responsible in 2(5%) of cases. Anemia was present in 28 patients(70%). ESR was elevated in 75% cases & repeat ESR done after 2 weeks was normal in all cases.Blood culture was positive in 7.5% cases,which grew Streptococcus Pneumonia. Chest X ray showed hydrothorax in 38(95%) & hydropneumothorax in 2(5%) patients. Ultrasonography of the chest revealed multiloculated empyema in 27, unloculated collection in 10(25%).One patient had a pneumatocele. Diagnostic pleurocentesis showed pus in 35 pts & 5 had seropurulent collection. The reported rate of identifying infective organisms in pleural fluid was 22.5%, commonest organism being Staphylococcus Aureus. . Most of the cultures were resistant to Ampicillin, Penicillin & Gentamicin. Staphylococcus aureus was sensitive to Vancomycin (100%), Cefuroxime (75%), Cloxacillin (50%).Patients who were culture positive were younger (2.05 yrs vs 5.04 yrs), were more malnourished (44.4% vs 38.7%,) had higher complication (1/9 vs 2/31), and decortication rate(3/9 vs 6/31) when compared to the culture negative group. One patient in the culture negative group died. More than 50% of cases required surgical management at time of admission. 19(47.5%) cases had primary VATS & 3(7.5%) required secondary VATS due to poor lung expansion & persisting fever with loculations. 9 patients(22.5%) required open thoracotomy . Pus drained ranged from 50 ml to 1000 ml. Lung expansion occurred at an average of 7 -12 days in most patients. 1 patient died due to severe sepsis. All 39 patients were followed up after discharge at 1 & 3 months.35 patients had good lung expansion,3 had pleural thickening & 1 had persistent collapse .Most of the patients were asymptomatic & had good lung expansion.

	Tube Thoracoscopic	Thoracotomy Debridement
The average number of ICD days	16 days	8 days
The average length of hospital stay	18 days	10 days

DISCUSSION

This study in children at a tertiary center found that empyema thoracis is more common in younger patients. A higher incidence of empyema has been reported in undernourished children(1,2).

In our study, the average number of ICD days was 16 with tube thoracotomy and 8 for thoracoscopic debridement.The average length of hospital stay was 18 days with tube thoracostomy as against 10 days with VATS .

The management of primary empyema continues to be controversial.In stage 1 empyema with no loculations ,antibiotics with tube thoracostomy would suffice,but in stage 2 empyema there is a changing trend towards VATS in a tertiary care hospital.

VATS was first used for treating empyema in children in 1993,as resue measure following failure of therapy with antibiotics & closed chest tube drainage(3).Since then, it has been suggested as primary therapy for parapneumonic empyema in children(4).

Early VATS provides an effective,singular procedure that combines cessation of the progression of the parapneumonic process,removes the infected pleural material,allows maximal lung expansion and function, with reduced pain and morbidity and a shortened hospital stay(5).Complicated empyemas will require thoracotomy and decortications.

Cohen et al(6) compared outcomes following the introduction of primary VATS in 21 children with atleast stage 2 empyema with a historical control group treated by chest drainage alone.There was a significant reduction in days in hospital (7.4 vs 15.4) and chest tube drainage(4 vs 10.2) in the VATS group.Furthermore,39% of patients treated with chest drain only required further surgical intervention ,compared with none with VATS group,suggesting that VATS is superior to chest drain alone.Chen et al,Karaman et al ,Eroglu ,Kercher et al, Doski et al(7) have all reported that early thoracoscopic debridement is effective and avoids need for surgical intervention.

Emphasis should be laid on minimizing the duration of hospital stay to bring down expenditure,psychological stress and more importantly nosocomial infections due to multidrug resistant organisms.Thus for appropriate patients, when facilities for VATS are available, VATS should be preferred.

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A PRETERM NEONATE WITH ECTODERMAL DYSPLASIA, CLEFT PALATE AND ENTROPION-A CASE REPORT

Dr. Gayathri H A, Dr. Prarthna V Bhardwaj

A 3hr old preterm female baby, born out of a non-consanguineous marriage, of birth weight 1.75kg delivered via naturalis, presented to us for low birth weight care. Baby had large areas on denuded skin involving the anterior aspect of the legs and forearms. Physical examination of the scalp revealed alopecia with crusting and scaling . Her fingernails and toe nails were not well developed. Both eyelid margins were turned inwards (entropion) and eyelashes were absent. Eyebrows were also absent. She had a widened nasal bridge and low set ears. The lower lip was found to be more prominent than the upper lip. (fig 1) Baby was found to have a cleft palate. Examination of the cardiovascular system revealed a systolic murmur.

Infant's mother denied febrile illness and usage of any drugs during pregnancy. She underwent regular antenatal check-ups. The mother was diagnosed with oligohydramnios prior to delivery of the baby. There is no similar case in the family.

An ECHO of the baby revealed a Patent DuctusArteriosus measuring 2mm. The baby was diagnosed with Hay-Wells syndrome without ankyloblepharon based on the clinical manifestations. Over the next 2 weeks, baby developed multiple episodes of sepsis as evident from the positive septic work up and the persistence of the umbilical stump at the end of 15 days, which required intravenous antibiotics. The neonate remained clinically stable. Fluids and broad spectrum antibiotics were administered intravenously. The denuded skin has slightly improved with emollients. She was discharged at 17 days of life and is doing well on follow up.

Fig-1

DISCUSSION

Hay Well's Syndrome was first described in 1976[1] as an autosomal dominant condition. This syndrome affects both males and females of all ethnic backgrounds. It is not known exactly how often it occurs. It is characterized by Ankyloblepharon, ectodermal dysplasia and cleft palate with or without cleft lip. Ectodermal defects include hair loss, absent or dystrophic nails, inadequate perspiration, widely spaced teeth and palmoplantarkeratoderma. Other associated anomalies include lacrimal duct atresia, supernumerary nipples, syndactyly and

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auricular deformities. It is rarely, associated with cardiac defects like Ventricular Septal defect or Patent ductusarteriosus. One study has suggested the association of this syndrome with genitourinary abnormalities and renal failure. [2] Intelligence is usually normal. Although these patients have a partial capacity to produce sweat from fewer glands so that hyperthermia is not a serious threat, heat intolerance is common.Since Hay-Wells syndrome is present from birth, diagnosis is based on the physical appearance of an infant or young child.[1] Various reports have been published describing newborn infants with Hay-Wells syndrome who were erroneously diagnosed with epidermolysisbullosa due to the presence of erythroderma and extensive areas of erosion. In association with the classic characteristics of this syndrome, skin erosion in the newborn infant and the recurrent scalp infection, are important signs that aid in the differential diagnosis with the other forms of ectodermal dysplasia.[3] One study has suggested that HWS is caused by heterozygous missense mutations in the p63 gene located at 3q27, which give rise to amino acid substitutions in the sterile alfa motif domain (SAM)[4,5]

As this syndrome is an inherited genetic disorder, treatment focuses on the symptoms present. Bacitracin ointment and mupirocin 2% ointment for skin lesions is required.[6] Surgery can be done to correct cleft palate. Speech therapy may be necessary because of cleft palate and/or missing teeth. Early ophthalmologic evaluation of lacrimal duct is required. Genetic counselling will be helpful for the individual and family affected by Hay-Wells syndrome. The importance of regular multidisciplinary follow up should also be stressed upon.

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PENTALOGY OF CANTRELL-A CASE REPORT

Dr. Madhusudan B S, Dr. Sudha Rudrappa, Dr. Pradeep N, Dr. Praveen kumar, Dr. Chinthan S

ABSTRACT :

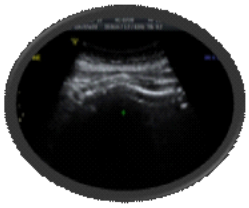
Pentalogy of Cantrell is a rare entity of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart which is due to deficient fusion of lateral folds of embryo. A 22 yr old mother delivered a still born baby with Pentalogy of Cantrell at Cheluvamba hospital Mysore.

INTRODUCTION :

The incidence has been estimated to be 1 in 65,000-100,000 live births(1,2). It is described as a deficiency of the anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, various congenital intracardiac abnormalities, and a defect of the lower sternum. This is causally heterogeneous. Cantrell suggested an embryological development failure of a segment of lateral mesoderm also called as "cephalic fold defect"(3).

CASE REPORT :

A 22 yr old lady, gravida two mother with 34 weeks of gestation presented with pain abdomen and bleeding per vagina. Ultrasonography showed multiple complex anomalies in fetus, including defect in anterior abdominal wall and sternum, with ectopia cordis and absent diaphragm. She delivered spontaneously a stillborn preterm baby, which had a large vcntral midline defect in thoracoabdominal wall with omphalocele and a part of liver protruding out through the midline gap along with a myelomeningocele, kyphotic deformity and club foot. Patient attenders did not give consent for autopsy and hence further evaluation could not be done.



Kyphotic Deformity

Omphalocele

Sternal Defect

Myelomeningocele and Club

DISCUSSION :

Pentalogy of Cantrell was first reported by Cantrell et al. (4) in 1958. J R Cantrell (1958) described the syndrome, consisiting of supraumbilical wall defect, defect of the lower sternum, deficiency of the anterior diaphragm, defect of the diaphragmatic pericardium and cardiac anomaly.

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The syndrome occurs with various degrees of severity from incomplete to severe expression with involvement of other organ systems. The etiology of the pentalogy is not well established. A widely-accepted theory, which was proposed by Cantrell et al.,(4) stated that developmental failure of the mesoderm in early embryonic life between 14 and 18 days of gestation results in a failure in the development of the transverse septum of the diaphragm, and of the ventromedial migration of the paired mesodermal folds of the upper abdomen.

Congenital defects of the sternum may vary from simple notching of the manubrium and irregularities in the shape of the xiphoid to absence of the entire sternum(5). Abdominal wall defects include omphalocoele, diastasis recti, epigastric hernia, umbilical hernia,and combined defects. The most common abdominal wall defect is omphalocoele(1). Deficiencies of the diaphragmatic pericardium and the anterior diaphragm are common defects. Sometimes, they are too small to be noted. Cardiac lesions may vary widely. Cantrell et al.(4) stated that congenital intracardiac anomalies are consistent elements of the pentalogy, with ventricular septal defect in every case (100%), atrial septal defect in 53%, pulmonary stenosis in 33%, tetralogy of Fallot in 20% and left ventricular diverticulum in 20%.In a collective review by Toyama(6), including 36 cases of pentalogy of Cantrell reported from 1772 to 1970, additional defects included head and facial deformities, meningocele, anencephaly, cleft lip, cleft palate, lung hypoplasia, adrenal aplasia, malrotation of the colon, hernia of the bowel into the pericardial cavity, undescended testicle, and deformities of finger and foot. Toyama WM (1972) described it in three categories

Category 1 : exact diagnosis with five defects present

Category 2 : probable diagnosis, with four defects (including intra cardiac and abdominal wall defects) present.

Category 3 : incomplete diagnosis, with combination of defects where sterna defect is always present.

Our case is an incomplete form of Pentalogy of Cantrell with anhydramnios, kyphotic deformity, intrabdominal cyst, myelomeningocele and club foot.

CONCLUSION :

Pentalogy of Cantrell is a spectrum of congenital anomalies, from fatal to nonfatal, that must therefore be adequately evaluated for appropriate prenatal counseling and postnatal management of individual cases. When the diagnosis pentalogy of Cantrell is suspected, a multidisciplinary approach is essential. These cases needs extensive corrective surgeries and they may develop cardio respiratory compromise following surgeries, especially in complete forms. Toyama (6) demonstrated a survival rate of 20% in this disorder including its variants and incomplete syndromes. The complete pentology has a poorer outcome, and the survival rate was only 5/59 (8.5%) in the report of Fernández et al.(7).

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A SIMPLE SCREENING-TOOL FOR FETAL- MALNUTRITION AT BIRTH-A COMPARATIVE STUDY OF CANS VERSUS OTHERS

Dr. Vishwanath Machakanur*, Dr. Sudha Rudrappa

ABSTRACT

OBJECTIVE : to assess the fetal malnutrition (FM) in term newborns using Clinical assessment of nutritional status (CANS) and comparing with other available methods.6

STUDY DESIGN: cross sectional study

METHOD :

225 full term neonates delivered at Cheluvamba Hospital attached to MMC & RI, Mysore were taken up for study,birth weight, length, mid arm circumference (MAC) and head circumference (HC) were recorded in the newborns. ponderal index (PI), wt/GA, MAC:HC ratio and MAC were calculated. Clinical assessment of nutritional status was done on the basis of CAN score and compared with other methods.

RESULTS :

Total 225 neonates studied with male:female ratio of 1.1 :1. CANS found 45.7% (103) neonates with FM. Wt/GA was more sensitive (98.06) and correlating statistically (p=0.000) with FM detected by CANS. FM detected by PI, MAC:HC ratio and MAC were less correlating and less sensitive to that of CANS (sensitivities = 86.40%, 51.45%, 50.48% respectively).

CONCLUSION:

CANS being a simple clinical-tool, can detect FM without aid of any sophisticated-equipment in term-neonates which are missed by other methods.

CANS can be used as a screening-tool in resource-limited settings of India effectively.

KEY WORDS:

CANS, fetal malnutrition, ponderal index, IUGR, SGA.

The incidence of low birth weight (<2500 grams) is still high in India (30%), as compared to the developed countries (5-7%). It is critical to screen and intervene early because of its high mortality and long term sequelae in neonates. Fetal malnutrition (FM) is a clinical state of obvious intrauterine loss or failure to acquire normal amounts of subcutaneous fat and muscles (first described by Scott and Usher).

METHOD:

This study was done over one month period of June 2013 and included 225 full term neonates, delivered at Cheluvamba Hospital attached to MMC&RI, Mysore.

- **Inclusion criteria:**
- Liveborn singleton neonates with >35 weeks gestational age (GA)
- Known GA (LMP, new Ballard score or Obstetrical ultrasound if done)
- No major congenital malformations

All babies born in the study period were weighed with electronic weighing machine and weight was recorded. Other anthropometric assessments were done at

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24-48 hour of life. Length of each baby was measured using infantometer. Mid arm circumference (MAC) was measured with the help of a non stretchable tape on left arm. Head circumference was recorded. CAN scoring was done for each baby.

Based on standard intrauterine-growth-curves (Wt/GA), neonates were classified into small and appropriate for gestational age (SGA, AGA respectively). Weght for gestational age (wt/GA) was estimated for each baby in terms of SGA or AGA using standard intrauterine growth curves. Ponderal index was calculated using formula [weight(grams)/Length(cm)3 ×100]. Values of <2.2 were taken as intrauterine growth retardation (IUGR). MAC/HC ratio <0.27 was taken as FM. MAC value <8.6cm was taken as FM. CANS (FM if <25 score) was considered standard and compared with other methods.

TABLE 1: The Nine Signs for Clinical Assessment of Nutritional (CAN) Status in the Newborn (Fig. l)(10).

1. **Hair**
   Large amount, smooth, silky, easily groomed (4). Thinner, some straight, "staring" hair (3). Still thinner, more straight, "staring" hair which does not respond to brushing(2). Straight "staring" hair with depigmented stripe (flag sign)(l).
2. **Cheeks**
   Progression from full buccal pads and round face(4), to significantly reduced bucal fat with narrow, flat face(l).
3. **Neck and Chin**
   Double or triple chin fat fold, neck not evident (4); to thin chin. No fat fold, neck with loose, wrinkled skin, very evident (1).
4. **Arms**
   Full, round, cannot elicit "accordion" folds or lift folds of skin from elbow or tricep area (4); to a striking "accordion" folding of lower arm, elicited when examiner's thumb and fingers of the left hand grasp the arm just below the elbow of the baby and thumb and fingers of the examiners right hand circling the wrist of the baby are moved towards each other, skin is loose and easily grapsed and pulled away from the elbow.
5. **Legs -Like arms.**
6. **Back**
   Difficult to grasp and lift skin in the interscapular area(4); to skin loose, easily lifted in a thin fold from the interscapular area (l).
7. **Buttocks**
   Full round gluteal fat pads (4); to virtually no evident gluteal fat and skin of the buttocks and upper posterior high loose and deeply wrinkled (l).
8. **Chest**
   Full, round, ribs not seen(4); to progressively prominence of the ribs with obvious loss of intercostal tissues(l).
9. **Abdomen**
   Full, round, no loose skin(4); to distended or scaphoid, but with very loose skin, easily lifted, wrinkled and "accordion" folds demonstrable.

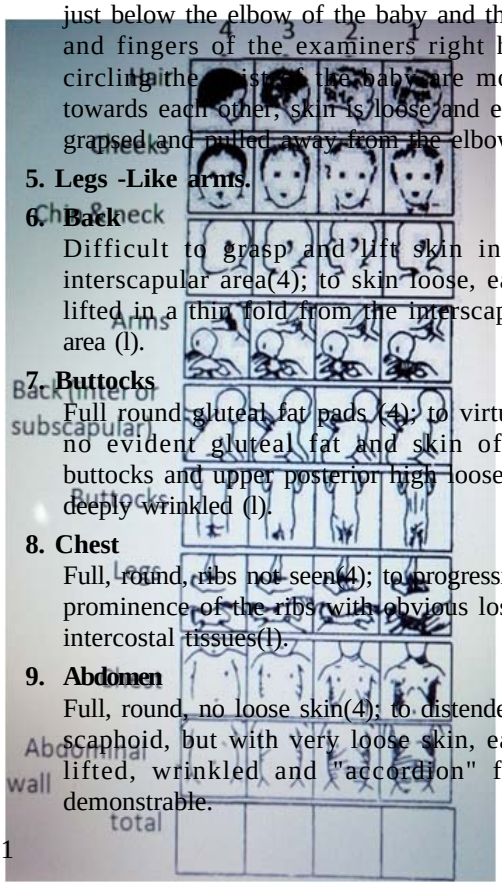

Table 1: comparison of CANS with Wt/GA.

		CANS		Total	P value
		FM	No FM		
Wt/GA	SGA	101(98%)	2(1.6%)	103	0.000
	AGA	2 (2%)	120 (98.4%)	122	
Total	103(100%)	122(100%)	225(100%)		

Table 2: comparison of CANS with PI

		CANS		Total	P value
		FM	No FM		
PI	IUGR	89 (86.4%)	2(1.6%)	91	0.000
	No IUGR	14 (13.6%)	120 (98.4%)	134	
Total		103(100%)	122(100%)	225(100%)	

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Table 3: comparison of CANS with MAC:HC ratio.

		CANS		Total	P value
		FM	No FM		
MAC/HC radio	FM	53 (51.5%)	3(2.5%)	56	0.000
	No FM	50 (48.5%)	119 (97.5%)	169	
Total	103(100%)	122(100%)	225(100%)		

RESULTS :

Out of 225 babies with sex ratio of 1.1:1, CAN scoring identified 103 babies (45.7%) as having fetal malnutrition (FM) and 122 (54.3%) as not with FM. Babies analyzed with wt/GA using standard charts, two were not identified as having small for gestational age (SGA) which were identified as having FM by CANS. Ponderal index (PI) could identify only 89 babies as having IUGR out of babies who were screened to be having FM by CANS. MAC: HC ratio identified only 53(51.5%) babies as having FM out of 103 babies of FM screened by CANS. MAC found only 52(50.48%) babies as having FM out of 103 babies with FM screened by CANS.

If we considered CANS as a standard tool to screen for FM, all other methods could not overtake or equate the fetal malnutrition detection rate of CANS.

Sensitivity of other methods, with CANS as standard were 86.4% for PI, 51.445% for MAC:HC ratio and 50.48% for MAC. Out of them Wt/GA was more sensitive to detect FM in trerms of SGA and correlated well with CAN score wth statistical significance ($p<0.01$).

CONCLUSION:

Hence, CAN Scoring being a simple clinical-tool, can detect FM without aid of any sophisticated-equipment in term-neonates which are missed by other

methods and can be used as a screening-tool in resource-limited settings of India effectively. Limitations of our study were: a small sample size and needed further larger sample size. Risk factors for IUGR were not included in the study, which may contribute to the conclusions.

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JARCHO-LEVIN SYNDROME WITH SPRENGEL'S SHOULDER DEFORMITY-A CASE REPORT

Dr. Jawhar E.A, Dr. Sachin Miraj, Dr. Basanth Kumar, Dr. Suresh Babu P. S,

INTRODUCTION

Jarcho- Levin syndrome is an autosomal dominant or recessive trait affecting segments of vertebra and anomalies of ribs.Clinically they present as chest wall deformities, short neck, short trunk and scoliosis. Jarcho-Levin syndrome has two subtypes 1) spondylothoracicdysostosis [STD] and 2) spondylocostaldyostosis [SCD]1.

CASE REPORT

A 5 month old female child,3rd born to a non-consanguinously married couple presented with respiratory distress.On examination child had short neck,low hair line,hemangioma on the nape of neck,low set ears,pectuscarinatum,left nipple at higher level,abdominal protuberance,lax skin,left scapula at higher level and scoliosis.

Chest X Ray showed left scapula at a higher level,scoliosis of thoracic spine,fused upper ribs on left side and thoracic hemivertebrae.USG abdomen showed left hydronephrosis.ECHO was normal.

Patient was treated with antibiotics and other supportive measures . Child improved and discharged on 7th day of admission.

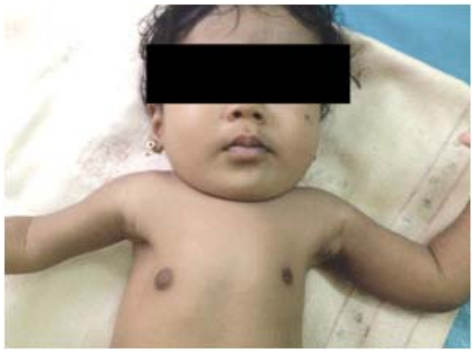

Fig 1 : short neck ,pectuscarinatum ,left nipple at higher level

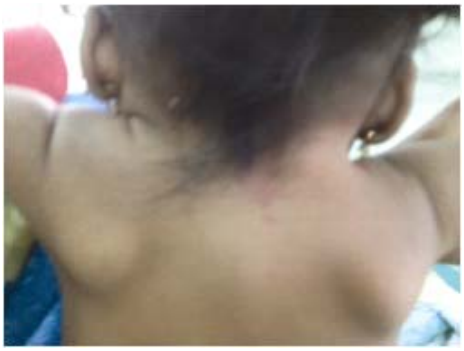

Fig 2 : low hair line,haemangioma at nape of neck,elevated left scapula

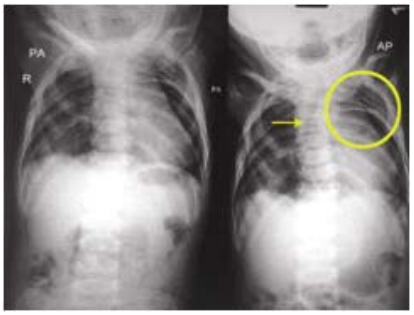

Fig 3 : scoliosis of thoracic spine, fused upper ribs on left side and thoracic hemivertebra

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DISCUSSION

Jarcho and Levin described this disorder in 19382.This syndrome described a spectrum of radiological and skeletal abnormalities, rib deformities and short trunk dwarfism.Solomon et al subdivided patients into 2 clinical phenotypes: spondylothoracicdysostosis (STD) and spondylocostaldysostosis (SCD).STD has bilateral fusion of ribs at costovertebral joints ,segmentation and formation defects of vertebra throughout the spine giving classical 'crab- like' or 'fan-like' appearance of the thorax.They are prone for pneumonia , restrictive lung disease with pulmonary hypertension and congestive cardiac failure. SCD is characterised by intrinsic rib anomalies such as broadening,bifurcation and assymetric fusion ,usually without severe thoracic impairment and better prognosis.Occasional abnormalities associated with Jarcho-Levin syndrome is cleft palate,hernias,hydronephrosis and neural tube defects.[3]

Association of Jarcho-Levin syndrome with Sprengel's shoulder deformity is rarely reported in literature[4].In this patient left scapula was at higher level.The non-descent of scapula might be due to segmentation arrest of somites resulted in Sprengel'sdeformity.Notch signalling pathway has important role in the segmentation and somitogenesis[5].

These patients are prone for recurrent pulmonary infection and respiratory insufficiency secondary to the small thoracic volume.These children should be monitored for growth &development,respiratory function and spinal curvature[4].Chest Physiotherapy has an important role in conservative management. Surgical interventions might be required in severe scoliosis.

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Represented Rajiv Gandhi University of Health Sciences in the Review meeting on Preventive Health programme in Geneva Switzerland held on May 19th to 25th Participated on Dengue Fever, Air Pollution.

He also participated in childrens Rights on environmental health organised by Human Rights Commission of Head quarters of United Nation.

ENDOBRONCHIAL TUBERCULOSIS : A DIAGNOSTIC DIALEMMA

Dr. Kavitha K, Dr. Mruyhunjaya. S, Dr. Gayathri Hemanth Aradhya *

ABSTRACT

Endobronchial tuberculosis (EBTB) is the tubercular infection of the tracheo-bronchial tree. It is a relatively uncommon detected manifestation of a common disease like tuberculosis and its incidence varies from 10% to 40% in patients with active tuberculosis and usually described in young individuals (1).

KEY WORDS:

Chronic cough, Endobronchial tuberculosis, mediastinal shift.

INTRODUCTION:

Endobronchial tuberculosis a special type of pulmonary tuberculosis, with increasing incidence. The tracheobronchial stenosis may cause intractable tuberculosis and make patients become chronic infectious source of tuberculosis, or may even cause pulmonary complications and result in death.(2)

Baby S A 1yr old boy resident of Davangere presented to out patient department with history of cough(non spasmodic) and hurried breathing for 3months, fever for 6days and with significant weight loss of 1-2Kg in last 3months, No history suggestive of foreign body aspiration.On examination there was increased respiratory rate(40-50/min) nomediastinal shift,impaired note heard on percussion over left lung field,On auscultation there was diminished air entry in the left lung field and occasional rhonchi in the left lung.

Chest x-ray revealed hyperinflation of left lung,(fig 1)with these findings Foreign body aspiration in the left main bronchus was diagnosed Provisionally. PPD reaction was measuring 10x8mm.With provisional diagnosis of foreign body in left main bronchus, diagnostic Bronchoscopy was done which revealed bronchial mucosa projecting into left main bronchus obstructing 75% of left main bronchus

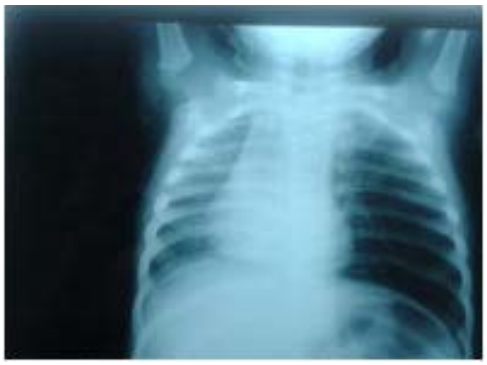

fig 1

CT Thorax :

Ball valve typeof obstruction at the left main bronchus due to granulation tissue secondary to mediastinal nodes,favouring the diagnosis of Pulmonary tuberculosis. (fig.2)

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Histopathology Of Mass: showed infiltrative granulomatous inflammatory lesion suggestive of Tuberculosis.

TREATMENT :

In the view of Endobronchial Tuberculosis Anti tubercular therapy Category 1 of RNTCP was started. Follow up for 6 months revealed good weight gain, decrease in cough and improvement in general condition.

DISCUSSION:

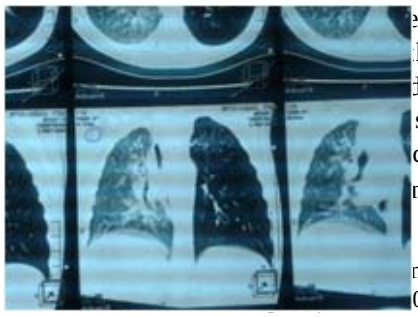
The clinical manifestations of EBTB may be acute, insidious or delayed with non-specific chest manifestations while modern treatment has led in overall decline of EBTB. Currently, its incidence may be underestimated since diagnostic bronchoscopy is not performed on every patient with tuberculosis. The pathogenesis of EBTB is not yet fully established. However, proposed mechanisms include direct implantation of tubercle bacilli into the bronchus from an adjacent pulmonary parenchymal lesion, direct airway infiltration from an adjacent tuberculous mediastinal lymph node, erosion and protrusion of an intrath oracictuberculous lymph node into bronchus, hematogenous spread and extension to the peri-bronchial region by lymphatic drainage.(3)

The clinical course of EBTB is variable because not only are there several possible pathogenetic mechanisms, but the interaction between the effect of mycobacteria, host immunity, and anti-tuberculous drugs is complex, and any variation in these three factors may result in an altered course.(4)

The early clinical manifestations of childhood ETB are often nonspecific. Although irritable cough accompanied with localized wheezing may be the important features of Endobronchial tuberculosis, they are not frequently seen. Chest X-ray and even CT often show normal results in the early stage.(5).Clinically, ETB is often underdiagnozed, or misdiagnosed as bronchitis, bronchial asthma, cough variant asthma, foerign body or bronchiectasis. Delayed treatment may result in tracheobronchial stenosis, systemic tuberculosis and posing serious effect on child's health and development.

CONCLUSION:

The eradication of Mycobacterium tuberculosis and the prevention of tracheobronchial stenosis are two most substantial treatment goals. To get the treatment goals, the diagnosis must be



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NEONATAL VARICELLA- A CASE REPORT

Dr. Archana MV, Dr. K. Shreedhara Avabratha*, Dr. Lokesha R

ABSTRACT:

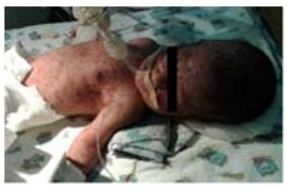
Neonatal varicella is caused due to peri-natal infection of the neonate. Clinical presentation may be mild to severe. Upto 31% of neonates with severe disease may succumb to it especially when maternal varicella begins 5 days or less before delivery or up to 2 days after delivery. A case of neonatal varicella developed on 10th day of life with a fatal outcome is discussed here with a brief review of literature to highlight the need for early initiation of aggressive treatment and importance of prophylaxis.

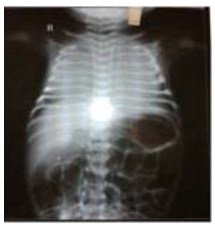
INTRODUCTION:

Neonatal varicella is rarely encountered(incidence of 2-6 per 100 000 live births per year[1]) and treated even less. Upto 31% of neonates with severe disease may succumb to it especially when maternal varicella begins 5 days or less before delivery or up to 2 days after delivery[2]. Clinical presentation is variable, ranging from a mild illness resembling chickenpox in older children to a disseminated disease like varicella pneumonia, hepatitis and meningoencephalitis[3]. Treatment involves varicella zoster immunoglobulin, acyclovir, isolation and supportive care. We report a case of neonatal varicella developed on 10th day of life with a fatal outcome with a brief review of literature.

CASE:

A 15 day old male baby was referred with varicella for further management. The baby was born full term to a 22 years primigravida mother by normal vaginal delivery at term in a hospital. The birth weight was 3.5kg. Mother on the day of delivery had developed chickenpox. Mother and baby were isolated for 5days and later were discharged home. Baby developed fever and erythematous papulovesicular rashes on the body on 10th day of life, and treated in a hospital with IV acyclovir and antibiotics for 5 days. As the baby developed respiratory distress and convulsions, he was referred. On examination he had tachypnea, poor peripheral perfusion, was drowsy with intermittent jerky movements of limbs and generalized vesicular rashes with few papules, ulcerated and crusted lesions. Investigations showed leucopenia with shift to left, moderate thrombocytopenia, with elevated CRP and liver enzymes. Chest X ray showed bilateral nonhomogenous opacities. Baby needed mechanical ventilation and anticonvulsants. Intravenous acyclovir and antibiotics were continued. Considering multi-organ involvement varicella zoster immunoglobulin was given to reduce the severity. In spite of all measures baby succumbed to the illness after 4 days.





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DISCUSSION:

Neonatal varicella is distinct from congenital varicella caused due to the intrauterine infection of the foetus. Neonatal varicella on the other hand is caused due to peri-natal infection of the neonate. The risk of infection and fatality rates are significantly high because of insufficient time for the development of maternal IgG and lack of passively transferred immunoglobulin protection in newborn and also the infant's cell-mediated immune response may not be enough to prevent the viremia.[4].

Any newborn with significant exposure to varicella zoster should be offered VariZIG (varicella zoster immunoglobulin) as soon as possible[5]. If VariZIG is not available and more than 96hous elapsed since exposure, one should consider giving IVIG or prophylactic acyclovir. A mother with active varicella lesions must be isolated. Treatment of neonatal varicella includes Intravenous acyclovir for 10 days and supportive/ symptomatic care as indicated, along with additional antibiotics if superimposed bacterial infection is suspected. There is no evidence VZIG modifies established varicella-zoster infections[6].

CONCLUSION:

Recognition of skin lesions, prompt diagnosis and treatment of pregnant women with varicella and neonatal prophylaxis are necessary to prevent neonatal varicella. The case report highlights the need for early initiation of aggressive treatment and importance of prophylaxis.

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SPECTRUM: STURGE-WEBER SYNDROME

Dr. K. Praveen Kumar*, Dr. Sachinmiraj, Dr. Ramesh.H, Dr. Ashwini.R.C, Dr. C. R. Banapurmath.

ABSTRACT

Sturge-Weber syndrome (SWS) is a neurocutaneous disorder, also called encephalotrigeminalangiomatosiswith angiomas that involve the skin of the faceandleptomeninges. The classical triad consists of -a facial cutaneous venous dilation, also referred to as a nevus flammeus or port-wine stain (PWS), leptomeningeal angioma and glaucoma.

We report 2 cases of SWS with varied presentations, one with classical triad of SWS and other with only central nervous system(CNS) involvement with no cutaneous or ocular manifestations, signifying that the diagnosis of SWS is not always straightforward.

KEY WORDS : Sturge-Weber syndrome, leptomeningealangioma, port-wine stain.

INTRODUCTION:

Incidence of SWS is 1 in 50000 live births. SWS is referred to as complete when bothfacial and central nervous system(CNS)angiomas are present, and incomplete when only the face or CNS is affected.

The Roach Scale is used for classification, as follows [3,5] :

- Type I - Facial and leptomenin-gealangiomas[LA]; patient may have glaucoma
- Type II - Facial angioma alone (no CNS involvement); patient may have glaucoma
- Type III - Isolated LA (with only CNS involvement); usually no glaucoma.

SWS is caused by residual embryonal blood vessels and their secondary effects on surrounding brain tissue. A vascular plexus develops around the cephalic portion of the neural tube, under ectoderm destined to become facial skin. Normally, this vascular plexus forms in the sixth week and regresses around the ninth week of gestation. Failure of this normal regression results in residual vascular tissue which forms the angiomataof theleptomeninges, face, and ipsilateral eye.[4]

A "vascular steal phenomenon" may develop around the angiomaresulting in cortical ischemia. Recurrent seizures, status epilepticus, intractable seizures, and recurrent vascular events may aggravate this steal further with an increase in cortical ischemia resulting in progressive calcification, gliosis, and atrophy which in turn increase the chance of seizures and neurologic deterioration.[10]

CASE REPORT :

Case 1 : A 9 year old boy, 5th issue of a non-consanguineous marriage, presented with acute history of headache, pain and redness in right eye, fever and one episode of convulsion. The child had past history suggestive of multiple focal seizures beginning at the age of 9 months and he is on antiepileptic medication till date.

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On examination vitals were stable and head to toe examination revealed port-wine stain(PWS) over right upper half of face. In view of these features, diagnosis of SWS was thought and further investigated. Completeophthalmic examination revealed increased intra ocular pressure(IOP) of 28mm of Hg in the right eye. CT brain showed right occipital tram track calcification. All the features were consistent with SWS type I. Child was managed initially with anticonvulsants for seizure control. Medical management for glaucoma with topical Timolol and Latanoprost eye drops,failed to decrease his IOP. Child underwent surgical correction (Trabaculotomy) following which his intraocular pressure is under control.

Case 2 : A 9 month old boy born to a non-consanguineous marriage presented with delayed attainment of age appropriate milestones and myoclonic seizures with no significantbirth and family history. On general physical examination there were no neurocutaneous markers. Ophthalmic examination was normal. MRI brain showed hemiatrophy of right cerebral hemisphere with leptomeningeal enhancement and gliosis of posterior parito-occipital region. Based on the neuroimaging study and the clinical profile features were consistent with diagnosis of SWS type-III(only CNS involvement with no port-wine stain).Seizures were controlled with two anticonvulsants. On follow up at 12m, 15m, and 18m child is seizure free and is gradually attaining the milestones.

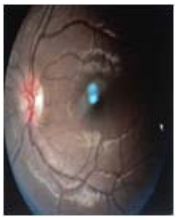
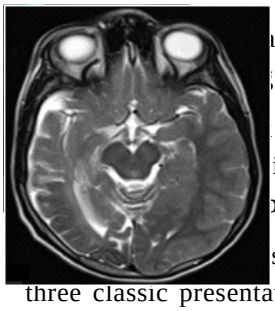


Fig 1: PWS over right half of face.
Fig 2: CT brain - Tram-track Calcification.
Fig 3: Fundus-Glaucoma

Fig 4 : Child with no PWS
Fig 5 : MRI brain-Rt cerebral atrophy and gliosis In parieto occipital area.

Discussion:



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se the child had all the three classic presentation of SWS- cutaneous manifestation as PWS, ophthalmic manifestation as glaucoma and CNS manifestation as seizures.

Special mention in this case was about ophthalmic involvement as glaucoma.

Glaucoma has been estimated to occur in 30-71% of patients in SWS[6]. Secondary glaucoma may present at any age, although early onset is the rule, with approximately 60% of glaucomas presenting at birth or in early infancy and another 30% presenting during childhood. The median ages reported

for onset of visual symptoms related to secondary retinal changes range from age 8-20 years.

It is produced by mechanical obstruction of the angle of the eye, elevated episcleral venous pressure, or hypersecretion of fluid by either the choroidalhemangioma or ciliary body. The anterior chamber angle abnormality is consistently seen in the infantile glaucoma cases in SWS, while increased episcleral venous pressure may have a key role in late-onset glaucoma cases in SWS. Decreased vision and blindness result from untreated glaucoma, with increased IOP leading to optic nerve damage. An acceptable range of IOP is 10-22 mm Hg.

The glaucoma associated with SWS is a significant cause of morbidity because of its early onset and resistance to conventional forms of treatment.

In our case the child presented as late onset glaucoma at 9 years and was not responding to conventional forms of treatement, requiring surgical correction (Trabeculotomy) to control his intraocular pressure. Following surgical correction his IOP is under control and is symptom free with good vision.

In our second case the child presented with seizures and developmental delay with no cutaneous markers .

Not all children with facial angiomas and PWS have SWS.The risk of SWS with facial PWS is about 8%.Incidence of SWS without facial PWS is estimated to be about 13%. In incomplete SWS (type III, Roach Scale), CNS angiomas occur without cutaneous features; therefore, no suspicion of SWS arises until a seizure or other neurologic problem develops. Thus, the diagnosis of SWS is not always straightforward [1,11]. Related to the degree of neurologic involvement, developmental delay and mental retardation occur in 50-60% of patients with SWS; they are more likely to exist in patients with bilateral involvement.[2] In a report by Sujansky and Conradi, using data obtained through the Sturge-Weber Foundation,[8] overall developmental delay occurred in 97 (58%) of 168 patients with SWS. Early developmental delay, however, occurred in 71% of patients with seizures and in only 6% of those without seizures.

In our case the diagnosis of SWS was made based on the typical neuroimaging findings and the clinical presentation. MRI brain showed diffuse right cerebral atrophy more marked in the parieto-occipital region with gliosis. These imaging finding suggest features of SWS.With effective seizure control measures most children regain their milestones as in this child.

CONCLUSION

SWS is a neurocutaneous disorder with sporadic inheritance. It is associated with various morbidities. The diagnosis is not always straight forward, requires imaging studies for confirmation. Timely intervention and good pschosocial support is essential for the overall development of the child.

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ASPHYXIATING THORACIC DYSTROPHY- A RARE SKELETAL DYSPLASIA.

Dr. Mahesh Maralihalli. Dr. Mahantesh Matti

INTRODUCTION:

Asphyxiating thoracic dystrophy is also known as Jeune syndrome. It is inherited as autosomal recessive disorder.

It is clinically diagnosed by its characteristic clinical features such as elongated, narrow and small chest, short arms and legs and postaxial polydactyly in some cases. There may be associated renal problems which can cause chronic renal failure in later life. There can be associated cardiac, liver, retinal, intestinal problems. Characteristic radiological features are bell shaped thorax,horizontally placed ribs, pelvic bone changes include short squared iliac wings, trident appearance of acetabular margin, wineglass pelvis, hypoplastic phalanges of hands and feet with cone-shaped epiphyses, short and wide long bones limbs (1,2,3).

The Severity of clinical presentation varies due to genetic heterogenesity of the disorder. There can be early neonatal death due to severe respiratory distress in severe form to mild medical problems in those survive beyond childhood to adolescents.

CASE DETAILS:

This baby, 1st by birth order, born of non consanguineous marriage, was born by emergency caesarian section. Baby cried immediately after birth. Baby had tachypnea at birth, which settled after 4 days. Feeds were initially given via nasogastric tube and slowly breast feeding started as tachypnea settled. Baby was discharged after 7 days of hospital stay. At the time of discharge baby had RR 48/min, HR 145/min, blood pressure within normal limits, systemic examination was normal. Salient clinical features were Short limbs, small and narrow chest, postaxial polydactyly in both upper limbs. These clinical features suggest Asphyxiating thoracic dystrophy. Investigations revealed normal blood counts, septic work up including white blood count, CRP, blood culture were normal. Chest X ray showed features of thoracic dystrophy. Ultrasound of cranium and abdomen were normal.

Figure1: An infantogram and clinical picture of the baby with typical features of Asphyxiating thoracic dystrophy

DISCUSSION:

It was first described by Jeune in 1955(4). Its incidence estimated at 1 case per 100000-130000 live births (5). Various

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terms used to describe the same are Asphyxiating thoracic chondrodystrophy, Asphyxiating thoracic dysplasia, Chondroectodermal dysplasia-like syndrome, Infantile thoracic dystrophy, Jeune syndrome, Jeune thoracic dysplasia, Jeune thoracic dystrophy, Thoracic asphyxiant dystrophy.

Jeune syndrome is a rare potentially lethal autosomal recessive skeletal dysplasia with respiratory and renal problems(6,4). Significant mortaliy is seen in early neonatal period because of severe respiratory insufficiency. Associated renal problems include cystic dysplasia or nephronophthisis. These renal problems progress to chronic renal failure later in the life. Dysgenesis, cirrhosis or fibrosis are the associated liver problems. Retinitis pigmentosa is seen in some patients. Intestinal malabsorption is infrequently seen in these patients. They may have pancreatic insufficiency later in life.

This baby had narrow tubular chest, short limbs, postaxial polydactyly. Radiological features on infantogram were suggestive of Jeune syndrome.

The differential diagnoses include Achondrogenesis, Achondroplasia, Cartilage-Hair Hypoplasia, Ellis-van Creveld Syndrome, Hypophosphatasia, Thoracolaryngopelvic dysplasia, Sensenbrenner syndrome.

The genetic basis of Asphyxiating thoracic dystrophy involves the IFT80 (3q25.33), DYNC2H1 (11q22.3), WDR19 (4p14) and TTC21B (2q24.3) genes, each encoding an intraflagellar transport protein, which confirms that Jeune syndrome is a ciliopathy.

These individuals need to be periodically screened for respiratory, renal, liver, cardiac, pancreatic functions, ophthalmologic examination. Since it is a autosomal recessive disorder, parents need to be counselled regarding the chances of same being inherited in offsprings.

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INDIAN ACADEMY OF PEDIATRICS Karnataka State
Medicare Center, Karangalpady, Mangalore 575003 (India)
Society Reg No: EKM -S460-2006-2007

PEDIATRIC TRAVEL FELLOWSHIPS

The Indian Academy of Pediatrics Karnataka shall award 6 (six) paid Trainee Fellowships every year to the members of the IAPK for training in Pediatric Specialties anywhere in India

Rules for selection of candidates

1. Age no bar.
2. The applicant should categorically indicate that the training received by him/her will be of use to the Institution / in private practice and that he/she has a definite plan to utilize the training by setting up the concerned specialty.
3. The candidate should clearly state which centre he/she wish to visit and for what specialty.

   The candidate will have to directly approach the Institution where he/she wish to have training and give a provisional letter of acceptance from the Head of Department under whom he/she will work.
4. The training will be for a period of 4 weeks.

   The IAP will provide a total grant Rs. 10000 (Ten Thousand) each.
5. The grant money will be paid to the Trainee Fellow on submission of

   (i) copy of training completion certificate

   (ii) a brief report on the training received by him/her and

   (iii) statement of expenses
6. The paid Trainee Fellowship is not open to those who have been awarded once by the IAPK.
7. Please submit duly filled applications on or before April 30th to

Joint Secretary,
IAP Karnataka Branch,
Medicare Center, Karangalpady,
Mangalore 575003

APPLICATION FORM FOR TRAINEE FELLOWSHIP

1. Name of the Applicant :			
Date of Birth			
Sex			
IAPK Membership No			
Address:			
Pin Code :			
Mob:		Email :	
2. Qualifications			
Medical / Pediatric Qualification			
Name of the University		Qualifying Date	
3. Appointments held till date:			
Sr. No	Designation	Period	Teaching /Non-Teaching
4. Proposed Sub-specialty and Subject of Training :			
5. Name and Address of the Institution where training is desired (enclose a letter of acceptance by the training institution)			
Pin Code :			
Phone:			
6. Give justifications for the training sought (Brief 100-150 words)			
7. Two References with Phone and email.id:			
i)			
ii)			

(Signature of Applicant)

Place :

Date :