

Experience in Counselling Down's Syndrome

I read with great interest the recent communication titled "Issues in Counseling for Down syndrome"(1). The communication is very interesting and highlights the lacunae and deficiency in the knowledge of pediatricians in handling parents of this common syndrome. I recorded 17 children of recognizable malformations, 9 of whom were phenotypic Down syndrome (chromosomal analysis not done in any child) while working at a community level charitable trust organization in Agra (UP) between January to November 2004(2).

These nine children were seen prior by pediatricians and were more than 5 years of age. All came from families of lower socioeconomic class (rural and urban slums). Only 3 parents were aware of the diagnosis, all knew about the incurability of the condition and presence of mental retardation. Few were fed up with the recurrent chest infections in their children and demanded an explanation for this. None were aware of the likely medical problems, schooling issues, recurrence in future children and what to do about it. All were depressed about the mental retardation but some were happy that their child is very friendly and cheerful.

In the absence of a qualified medical geneticist in the town at the time, unwillingness of parents to go to other towns for specialized medical genetics services and available counseling services being

unaffordable to most families, the best I could do for them was to screen for medical problems and do the counseling myself. I tailored the issues specific to individual families, highlighted the need for vocational training in long term and discussed and motivated all parents regarding future pregnancies. I used established strategy of motivational interviewing to increase family's adherence to specific issues(3). I was disappointed to be only partially successful and felt a need for further training. The bulk load of children requiring counseling services comes to practicing Pediatricians first. It may not be feasible for every practicing pediatrician to undergo training courses in counseling after post-graduation due to different preoccupations. The best possible way seems to include a short-structured training module in genetic counseling during postgraduate training program in Pediatrics.

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Kawasaki Syndrome in Coastal India

In response to your editorial(1) we present our observations on 26 children diagnosed with typical Kawasaki Syndrome (KS) conforming to American Heart Association criteria in a coastal district of

South India between 1999 and 2006. All were referred, 30% (3/10) with probable diagnosis of KS before 2004 and 62.5% (10/16) thereafter. Mean age in years at presentation was 5 till 2004 and 3.5 thereafter with equal sex ratio contrary to male predominance reported worldwide. Though all children presented with fever, mean duration of

fever prior to referral were 8 and 6.5 days respectively in the two time frames. Unilateral cervical lymphadenopathy was seen in 92%; nonpurulent conjunctivitis and skin rash in 88%; oral mucosal changes in 85%. Of the 81% with desquamation, 71% developed periungual and perianal desquamation within 10 days of illness, similar to reported observations(1). Arthralgia was present in 42%. Cardiac complications were seen in 23% which included coronary artery dilatation in 3, coronary artery aneurysm in 2 all of which regressed in the follow-up, and congestive cardiac failure in 1 with unresolved mitral and tricuspid regurgitation in the five year follow-up. Elevated ESR and positive CRP were observed in 96% and 88% respectively. Thrombocytosis was seen within 10 days in 73%. Intravenous immunoglobulin given to 73% of children was well tolerated with one requiring two doses of 2 g/kg. All received high dose aspirin during the acute phase followed by low dose of 3-5 mg/kg/day for six weeks. There was no mortality.

On analysis of Indian case series totaling five or more(1,2), there seems to be a concentration of cases in coastal cities numbering to 206 (73%), against 75 from rest of India. The highest incidence of KS from United States is from Hawaii and within the continent from West Coast(3). Japan, Taiwan and Hong Kong account for most cases of KS in Asia. In contrast, Great Britain and Australia have very low incidence(4). A Washington study(5) in response to

three outbreaks of KS in close residential proximity to water bodies recommends further studies exploring the relation of Kawasaki disease occurring more in proximity to sea-shore!

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Tuberculosis Infection in BCG Vaccinated and Non-vaccinated Children

The paper on prevalence of tuberculosis (TB) infection according to BCG vaccination status by Pulickal and Fernandez is appreciated for the focus on childhood TB infection, especially in Kerala where it had not previously been explored(1). Several studies in other parts of India report lack of protection of TB infection by BCG vaccination(2). The protection reported in Kerala appears to be

artifact of analysis. Using 10 mm cut-off value for PPD response in non-vaccinated and 15 mm in vaccinated children deflates sensitivity of TB infection in the latter(2-5). Different cut-off values is inappropriate in school-age children in India(2-5). The recommendation is to ignore BCG status for assessing TB infection prevalence by PPD test(2,3,5).

The analysis should have started with determining the cut-off values appropriate for Palakkad region, by drawing superimposed graphs of frequency distributions of reaction sizes in both