



CrossMark

Highlights from this issue

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ABANDONMENT OF CHILDHOOD CANCER TREATMENT

Abandonment of childhood cancer treatment is a major contributor to treatment failure in low income countries and virtually unknown in high income countries. The reasons for this are complex. Njuguna and colleagues explore this in a descriptive study using semistructured questionnaires to interview families of childhood cancer patients diagnosed between 2007 and 2009 who had abandoned treatment at the Moi Teaching and Referral Hospital in Kenya. There were 222 children diagnosed with treatment outcome documented in 180. Of these 98 abandoned treatment. Fifty-three families were traceable and 46 agreed to be interviewed. Reasons for treatment abandonment were—financial difficulties (46%), inadequate access to health insurance (27%), transportation difficulties (23%)—the full list is in the paper. Most abandoned treatment after the first 3 months. Of the 46 children only 9 (20%) were still alive at the point of family interview, of which six looked healthy.

Change requires improved access to health insurance, financial or transportation support, parental education, psychosocial guidance and improved communication. The wider issue of why children do not receive treatment is discussed in an accompanying editorial. The challenge is that in many countries health care is a commodity for those who can afford it rather than a right. *See pages 609 and 605.*

ASSESSMENT AND MANAGEMENT OF ANXIETY DISORDERS IN CHILDREN AND ADOLESCENTS

Anxiety disorders are among the most common psychiatric conditions seen in young people with a significant potential adverse impact on educational achievement, family life and leisure activities. They often co-occur with other medical, psychiatric and behavioural disorders. Family history is common and longer term anxiety disorders are associated with an increased incidence of anxiety and depression in adult life. Cresswell and colleagues discuss the assessment and management—specific phobias, separation anxiety disorder, social anxiety disorder, panic disorder and agoraphobia. The evidence base and practicalities of cognitive behavioural therapy are discussed including low intensity interventions that have been proposed as a means to improve access. The risks and benefits of

pharmacological therapies are discussed with the authors advising caution about routine use. The review is comprehensive and helpful and essential reading for clinicians who regularly see children and adolescents with medical disorders where anxiety is a significant factor and intervention may impact. *See page 674.*

CARDIOVASCULAR DISEASE IN NOONAN SYNDROME

Noonan syndrome (autosomal dominant, variable penetrance) is the most common syndromic cause of congenital heart disease after trisomy 21. Incidence is 1 in 1000 to 1 in 2500 live births. Prendiville and colleagues report the spectrum of cardiac abnormalities and clinical course of a large cohort (n=203). Cardiac disease was present in 81% (N=237) including pulmonary stenosis (57%), ASD (32%) and hypertrophic cardiomyopathy (16%). In those with pulmonary stenosis (n=166) the majority had additional cardiac defects including ASD or hypertrophic cardiomyopathy or both. A genetic mutation of the RAS-MAPK signalling pathway was identified in 62% with specific phenotype/genotype associations for different cardiac defects. Outcome data is of interest with median age at last follow being 14 years. Surgery (n=35) or percutaneous balloon pulmonary valvuloplasty (PBVP, n=43) was required in 47% with pulmonary stenosis. Of those who underwent PBVP 65% required further intervention—either repeat PBVP or surgery. Of the 47 with hypertrophic cardiomyopathy more than 50% were diagnosed in the first year with additional cardiac lesions in 70%. Of the cases diagnosed in infancy 8/27 underwent spontaneous regression and 4/27 died, one from complications of preterm birth. The spectrum of conditions and reported outcomes is important for clinicians and families and will help facilitate counselling on outcome and prognosis. *See page 629.*

LONG QT MOLECULAR AUTOPSY IN SUDDEN INFANT DEATH SYNDROME

Long QT syndrome is a group of channelopathies with 13 known genotypes which predispose to sudden death due to ventricular tachycardia. These are investigated by 'molecular autopsy' in sudden death age 1–40 years. The role in sudden infant death is less clear where there are multiple potential risk factors. In this issue Glengarry and colleagues investigate this in an unselected and then selected cohort.

It is an interesting paper to work through. Rare genetic variants were commoner in the selected (5/31) than the unselected cohort (3/71). Not all the variants were clinically significant, although risk factors are a predictor. Further investigation is time consuming and requires family engagement. The authors suggest a diagnostic yield of 10% with appropriate case selection and suggest testing should be confined to—family history of previous sudden deaths, syncope, seizures or proven cardiac arrhythmias; absence of known risk factors, particularly bed sharing with possible overlaying; sudden collapse while awake; high quality autopsy and report exclude other possibilities. *See page 635.*

BIOLOGICAL THERAPIES IN PAEDIATRIC RHEUMATOLOGY

The last 15–20 years have seen major developments in the treatment of juvenile rheumatic diseases. Sen and colleagues, in a comprehensive review, update us on new biological therapies including the discussion of the mechanisms, indications, efficacy and toxicity. The goal of inactive disease with minimal disease associated damage is becoming a reality. With the potential need for long term treatment the importance of national and international registries, so that long term safety and efficacy can be monitored beyond clinical trials, is emphasised. *See page 679.*

IN F&N THIS MONTH

Cleft lip with or without palate can be diagnosed antenatally by ultrasound and may be classified as apparently isolated versus associated with other malformations. There is little data on the outcome of 'isolated' facial clefting. Burnell et al, by retrospective case note review, report the longer term outcome of such cases. 15/97 pregnancies were terminated. Follow up data was available for 77/81 live born infants. Major malformations were found in 15%—including familial clefting syndromes, trisomy 21, autistic spectrum disorders, brain malformations, fetal alcohol syndrome and Kabuki syndrome. A further 11/77 had additional anomalies of minor impact including cardiac anomalies and developmental delay. No child with clefting only had any additional anomalies. The authors rightly conclude that there should be a low threshold to investigate concerns in an infant with orofacial clefting. The term 'isolated' facial clefting should clearly be used with caution.