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# Highlights from this issue

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## PREVALENCE AND ASSOCIATED HARM OF ENGAGEMENT IN SELF-ASPHYXIAL BEHAVIOURS

Busse *et al* report a systematic review looking at prevalence of engagement in self-asphyxial (risk taking) behaviour ('choking game') and associated morbidity and mortality in children and young people up to age 20 years. Awareness of self-asphyxial behaviours was high, 36–91% across different settings. Median life time prevalence of ever engagement in young people is 7.4% based on cross sectional data from North America, France and Colombia. Fatal cases (99) have been reported in 10 different countries. Most occur when individuals engage in self-asphyxial behaviours on their own, and use ligaments to engage in the process. The public health implications of this data are complex and are discussed in the paper and accompanying editorial—Risk taking behaviour in adolescents; 'Chance only favors the prepared mind.' In essence the available evidence indicates that it is not enough for children and young people just to know about self-asphyxiating behaviours, they must also understand the social context of engaging in these behaviours, and that the decisions they make about them can have significant consequences. *See pages 1106 and 1115*

## MANAGEMENT OF DUCHENNE MUSCULAR DYSTROPHY

Duchenne muscular dystrophy is the commonest inherited neuromuscular disorder of children. Over the last 25 years life expectancy has doubled as a consequence of improvements in diagnosis, treatment and long term care. Strehle and colleagues review the recent scientific advances and their impact and potential impact on children with this devastating and life limiting condition. The importance and practicalities of early diagnosis are highlighted. There is a comprehensive consensus management guideline adopted by the National Institute of Clinical Excellence and comprehensive standards of care best delivered by a dedicated multidisciplinary team and covering the multiple body systems affected—muscular, respiratory, cardiac, gastrointestinal, skeletal, renal/urogenital and nervous. Corticosteroids should be given early and help to maintain

muscle strength and function. Multiple other drugs can be used and are listed. The research focus is on new therapies, the potential for gene therapy and the potential for cure and these are discussed in detail. 'Small molecule therapies' that interfere with specific gene mutations carried by subgroups of boy's with DMD appear to be the most promising. Gene therapy has the potential to impact but is still in its infancy despite many years of intensive research. In the meantime standardised conventional therapy and regular monitoring by a dedicated team of health-care specialists should remain the cornerstone of best management. *See page 1173*

## WHAT MATTERS TO CHILDREN WITH CFS/ME

Chronic fatigue syndrome/myalgic encephalomyelitis (CFS/ME) is common and disabling with a significant impact on mood and school. Like any other chronic illness well developed patient related outcomes measures (PROMs) are important to inform clinician's and researchers regarding best management. PROM development starts with a patient derived conceptual framework that defines the key outcomes to be measured. Parslow and colleagues explore the aspects of life and health outcomes that matter to children with CFS/ME. 25 children were interviewed (mean age 13 range 8–17, 14 female, 19 with their parents present) and three mothers participated in a focus group. Children identified four key themes (health outcome domains)—'symptoms' that fluctuated, which caused an unpredictable reduction on both 'physical activity' and 'social participation' all of which impacted on 'emotional well being'. These domains were influenced by both management and contextual factors. The relationship between healthcare and school was considered pivotal. The themes and interactions are illustrated (with examples) in figure one. The awareness of how children conceptualise CFS/ME is fundamental to effective management and should inform questions asked and topics discussed in the clinical setting. *See page 1141*

## PRESENTATION OF CHILDHOOD CANCERS TO A PAEDIATRIC SHARED CARE UNIT

The two week (suspected) cancer referral pathway is designed to reduce the time to

diagnosis although the impact in childhood is controversial. Roskin *et al* describe the pathways by which children with cancer presented to their shared care unit (93 children, 0–15 years, 2004–14). Only 2/93 were referred via the two week pathway. Most (62/93, 67%) of presentations were acute via immediate GP referral or self presentation to the emergency department, leukemia most often via GP referral and solid tumours by self referral. Others were referred mostly from general paediatric/specialist clinics. Median time from symptoms to presentation was 20 days (time from symptoms to diagnosis 31 days) although there was a large range—3 days to presentation, 17 days to diagnosis for leukemia; 12 days to presentation, 27 days to diagnosis for brain tumours; 107 days to presentation, 120 days to diagnosis for lymphoma. The two week pathways doesn't really contribute. The authors recommend that the role of the 2 week pathway in paediatric cancer care should be reviewed. *See page 1131*

## CHILDREN WITH ORAL CLEFTS AT RISK OF LOW ACHIEVEMENT IN SCHOOL

Children with isolated oral clefts are at greater risk of doing less well at school than unaffected peers although there is cross sectional data that suggests children may 'catch up' during adolescence. Wehby and colleagues report longitudinal data of academic achievement (586 children with oral clefts, 1873 unaffected classmates) from early elementary school through to high school. Achievement trajectories were stable for both groups. Children with oral clefts were significantly more likely to be classified into persistent low achievement trajectories even when adjusted for socioeconomic differences: OR 1.63 (95% confidence interval 1.23–2.16) for reading, OR 1.73 (95% confidence interval 1.19–2.31) for language and 1.45 (95% confidence interval 1.05–1.99) for mathematics. Predictors of low achievement were cleft palate only, adolescent mothers, low maternal education and less frequent use of prenatal care. These findings support the need for routine, early screening for academic deficits in this population and remind us that school factors are an important part of the clinical assessment when these cases present to clinic. *See page 1148*