Highlights from this issue

R Mark Beattie, *Editor in Chief*

IMPROVING RECOGNITION OF DUCHENNE MUSCULAR DYSTROPHY

Duchenne muscular dystrophy is a life limiting muscle wasting disorder typically presenting with delayed or disordered motor or speech development and muscle weakness. The last decades have seen improved standards of care and new therapeutic approaches–corticosteroid treatment, nocturnal ventilation and cardiac support with significantly improved clinical outcomes and life expectancy. However, the condition (X linked, 1 in 4000) tends to be diagnosed late which has a negative impact on the potential for genetic counselling and recruitment into clinical trials. Van Ruiten and colleagues look at the 'diagnostic process' in 20 children diagnosed over 10 years as a useful way of investigating the cause of (and potential strategies that can reduce) diagnostic delay. The results are of interest—age at first reported symptoms 32.5 (range 8–72) months, age at first engagement with a health professional 42.9 (range 10–90) months, creatine kinase levels checked at 50.1 (14–91) months, diagnosis confirmed at 51.7 (range 16–91) months. The age at diagnosis is less than reported previously although there are significant delays in presentation and investigation following presentation. The authors advocate screening as part of the two year developmental check and promote the mnemonic MUSCLE—Motor milestone delay, Unusual gait, Speech delay, CK ASAP Leads to Earlier diagnosis of DMD. The findings are discussed in an accompanying editorial. ***See pages 1074 and 1061.***

WHY DO PARENTS LITIGATE AND WHAT DOES IT DO FOR THE FAMILY

The annual cost to the NHS of litigation was more than £1 billion in 2012. A significant component of that was paid to claimants in brain damage at birth litigation. Lewis Rosenblaum—who has prepared about 5,000 reports looking at causation of brain damage and disability—gives his perspective based on his extensive experience of why parents litigate. It is interesting to work through and gives us a perspective on the issues that these families face and the importance of specifics. These include what (exactly) has happened to cause brain damage in their child, to stop this happening to other children, to ensure their child is properly cared for (including in adult life), to obtain retribution against perceived errors, to respond to pressures from family or other agencies and to deal with the unexpected discovery of possible fault. Clearly financial compensation is a major factor and relevant to all of the above. The issues listed are often difficult to address with families and this article reminds us of the importance of at least trying to address some of them—particularly what has happened to cause brain damage in their child—in order to help families comes to terms with and deal with the reality of caring for a disabled child long term. ***See page 1065.***

PROPRANOLOL FOR INFANTILE HAEMANGIOMA

Propranolol has become first line treatment for complex infantile haemangiomas. Solman and colleagues report their experience of treating 250 children (34% preterm) who completed at least 3 months of therapy. Indications included visual compromise (30.4%), airway obstruction (8.8%), feeding difficulty (8.4%), risk of permanent disfigurement (4.4%). Median age at start of treatment was 4.5 months, Median length of treatment was 11.8 months. 96% responded well to therapy–20 patients experienced re growth off treatment and 6 required propranolol to be re started. 38 (15.2%) patients experienced side effects—mostly mild—including wheeze, worsening of ulceration, sleep disturbance and diarrhoea requiring modification of treatment in 26. These data confirms that propranolol is safe and effective and appropriate to use in selected cases in whom there are complications of infantile haemangiomas. The paper includes detail on the indications, potential toxicity and practicalities of use—modified based on their experience—including a helpful treatment protocol. ***See page 1132.***

COMMON VISUAL PROBLEMS IN CHILDREN WITH DISABILITY

There is a high prevalence of visual problems in children with disability. Alison Salt and Jenefer Sargent review the aetiology including risk factors such as pre-term birth, cerebral palsy, learning difficulty, syndromal disorders and primary visual impairment. The authors offer a comprehensive approach to the identification, assessment and management highlighting the need for better screening and service provision for children with visual impairment and neurodisability. ***See page 1163.***

TOXIC SHOCK SYNDROME

Toxic shock syndrome (TSS) is an acute toxin-mediated illness caused by toxin-producing strains of staphylococcus aureus and streptococcus pyogenes. Risk factors include chicken pox, burns and tampon use. Adalat and colleagues report 49 children (November 2008–December 2009, BPSU surveillance data, UK) – overall incidence 0.38 per 100,000. This may be an underestimate as a consequence of under reporting, only the most severe phenotype being recognized and reported. 29 out of 49 were likely secondary to streptococcal infection (18 confirmed), 20 were secondary to staphylococcal disease (15 confirmed). Children with staphylococcal TSS were generally older. Most (78%) required intensive care support. Agents with antitoxin effects were used, although not in all; Clindamycin 67%, IVIG 20%. There were 8 deaths—all in the streptococcal group, none had received IVIG. The high proportion of streptococcal TSS and higher mortality are highlighted by the authors who emphasise the need for an agreed guideline to improve management of TSS in children. Nigel Curtis, in an accompanying editorial discusses TSS: under recognised and under treated? ***See pages 1078 and 1062.***

IN *E&P* THIS MONTH

Managing the adolescent with back pain

Non specific back pain is common in adolescence and can be difficult to assess. Cruikshank *et al* offer a first rate practical approach as part of the 15 minute consultation series. The authors emphasise the importance of a structured approach including a careful history (there is a table of red flags), musculoskeletal examination including pGALS (there is a table of red flags), management strategies including simple analgesia and physiotherapy and when to investigate further/refer for specialist advice. There is an excellent 'test your knowledge' series of case based extending matching questions to assess your knowledge at the end with detailed and helpful explanations for each of the different scenarios given.