NEUROMUSCULAR CONDITIONS

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Joint stability and limb function rely on the complex integra - tion of the musculoskeletal and neurological systems. Damage to either leads to one of several conditions linked only by the during the period of skeletal growth. Management is directed at helping the child cope with their disability , minimising further deterioration and maximising function. It is important to have an understanding of what the damage is and what the future holds (Table 44.14). Spina bifida and polio are classic lower motor neurone lesions, whereas cerebral palsy and head injuries a ff ect upper motor neurones and the higher centres. There are often other disabilities such as blindness, epilepsy and intellectual di ffi cul ties to consider. /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF In children, even if the initial insult to the neuromuscular system is non-progressive, the e ff ects of the insult change with growth. Damage at any level of the neuromuscular system leads to an alteration in tone and muscle imbalance associ ated with decreased control of movement. Abnormal muscle pull, particularly in combination with the e ff ects of gravity , alters bone growth, leading to deformity and joint contrac ture. Muscles are relatively weak and, with body growth and a weight increase, they are no longer strong enough to control a heavier limb, particularly when deformity means they are working a t a mechanical disadvantage. A multidisciplinary approach to management is essential. Physiotherapy and orthotic management may reduce the need for surgical intervention and postoperatively they ensure that the surgical benefits are maximised. In conditions such as Duchenne muscular d ystrophy there is substantial evidence for the benefits of certain surgical procedures; however, in other conditions (cerebral palsy) there are fewer such long-term validated studies. /uni25CF /uni25CF Guillaume Benjamin Amand Duchenne (Duchenne de Boulogne) held a hospital appointment. ment, muscle length and tendon excursion. This is easier to achieve in patients with a flaccid paralysis or low tone. The maintenance of muscle strength is also important. The use of splints, positioning techniques and seating and sleeping sys - tems is common with the aim of preventing fixed contractures. Surgery has a valuable role in the management of selected patients (Table 44.15). The surgeon must understand that altering ankle posture may a ff ect knee and hip posture/function and vice versa. - The patient m ust have the intellectual ability and motivation to recover fr om the surgical procedure. Some of the factors mentioned previously (Table 44.14) must be considered in any holistic approach to the patient. Summary box 44.19 Principles of treatment of neuromuscular conditions /uni25CF /uni25CF /uni25CF /uni25CF - -

TABLE 44.14 Factors to be considered in the assessment of a neuromuscular disability. Is the insult to the neurological system progressive or non- progressive? Is it located centrally or peripherally? Is it general or focal? Is it associated with other abnormalities or not? If the insult is not neurological, is it myopathic? TABLE 44.15 General types of surgical procedure that may be

considered in the management of a patient with a neuromuscular condition. Surgical procedure Lengthening of the muscle–tendon unit Tendon transfer Improves functional movement; rebalances muscle forces, after Release of joint contracture; correction of bony deformity Fuse/stabilise/relocate joints Neurological procedures: selective dorsal rhizotomy (SDR) intrathecal baclofen pumps (ITB) Leg equalisation procedures A neurological defect, whether progressive or not, may cause progressive deformity with skeletal growth A multidisciplinary approach is essential Primary therapy aims to maintain range of movement and prevent /f_i xed contractures with an emphasis on managing tone and position Surgery has a limited role in the management of neuromuscular conditions

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