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Acute paediatric paraplegia: A case series review

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ABSTRACT

Paediatric paraplegia resulting from spinal cord pathology of any cause is rare; hence prognostic information for children less than 16 years is limited. This case series review aims to ascertain all cases of paediatric paraplegia from 1997 to 2012 in the former Northern Region of England.

Methods: Children presenting with sudden paraplegia before the age of 16 years were multiply ascertained from databases in the regional paediatric neurology, neuroradiology, neuro-oncology and adult spinal injuries units. Data were obtained from retrospective case note review.

Results: A total of 44 cases (24 female) were identified. The incidence is estimated at 0.49 per 100,000 children under 16/year (95% confidence interval 0.41–0.57). Mean age of onset was 8.8 years and the most common aetiology was inflammatory. Twelve months post presentation, mortality was zero and a good outcome (defined as Gross Motor Function Classification System grades I or II) was seen in 66.6%. Motor outcome at 12 months was associated with the presence of bladder/bowel signs at presentation, previous viral illness and initial severity of paraplegia. Bladder signs at presentation were the strongest predictor of prognosis (OR for poor motor outcome 10.3). We were unable to demonstrate a relationship between aetiology and late outcome.

Conclusion: Paediatric paraplegia is rare. Mortality rates are low and 66.6% have a good outcome with fully or nearly independent walking. Bladder signs are the strongest predictor of prognosis.

1. Introduction

Paraplegia is a clinical syndrome of signs and symptoms reflecting spinal cord dysfunction, usually below a definable anatomical level. The four main aetiological groups in children are trauma (e.g. falls, or penetrating injury such as gunshot), vascular (including anterior spinal artery infarction), inflammatory (including both primary infection or abscess and parainfectious processes such as transverse myelitis and encephalomyelitis) and compression (tumour, syringomyelia).

The epidemiology of adult paraplegia has been well documented. It is estimated that 130,000 people worldwide suffer spinal cord injury (SCI) each year and an estimated 2–3

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million live with the associated disability.[1] Incidence peaks between the ages of 15–30 years and is highest in males primarily because of traumatic SCI sustained in road traffic accidents.[2–4] There is a second peak in incidence over 65 years due to falls, and non-traumatic age-related aetiologies such as tumour compression, spinal stenosis and vascular ischaemia.[4] It is commonly accepted that final neurological outcome is reached within one year of injury.[5,6] In adult traumatic SCI, complete injury (of both sensory and motor systems) accounts for approximately 50% of cases and is associated with a poorer prognosis and increased mortality.[1,5] Severity of neurological injury and the presence of multi-system trauma have been found to correlate with functional recovery.[5,7] Cervical cord injury is associated with a better functional recovery than thoracic cord injury, with restoration of walking and bladder function in 30% and 20% respectively compared with 10% restoration in both for thoracic cord injuries.[8] Thoracic SCI is typically associated with more severe trauma involving cardiopulmonary dysfunction and impaired blood supply to the watershed site of the spinal cord injured at that level.[8]

Paediatric paraplegia is rare and much less well documented, in a largely aetiology-specific literature. An Australian study identified that the two most common causes of acute flaccid paralysis were Guillian–Barré syndrome and transverse myelitis with Guillain–Barré syndrome accounting for 47% of presentations.[9] A nationwide cohort study from Taiwan showed that the overall incidence specifically of traumatic SCI in those aged under 18 years was 5.99 per 100,000 person-years (68% cervical). Incidence rose in adolescence and was commoner in males. Motor vehicle accidents were the most common cause.[10]

Brown et al. found that motor vehicle accidents were the most common cause of cervical traumatic SCI in young children and sporting injuries in adolescents.[11] As in adults, prognosis for recovery among children was associated with neurological status at presentation[10,12] and those with incomplete injuries recovered more functional independence.[13]

Some evidence suggests that children have a better neurological recovery than adults.[12] It is thought that adult co-morbidities such as hypertension and diabetes mellitus impede recovery. Additionally children may benefit from greater plasticity of the immature spinal cord.[14,15] However it is known that very young children tolerate SCI poorly.[16] The complications of SCI, including urinary dysfunction, spasticity, pain syndromes, respiratory insufficiency and psychological sequelae, are especially challenging for children and adolescents.[17] Furthermore children who suffer injury prior to their growth spurt are at an increased risk of developing scoliosis (97% compared with 53% sustaining SCI after puberty).[18]

A prospective population-based surveillance study by De Goede et al. showed that the annual incidence of transverse myelopathy (of inflammatory and vascular aetiologies) in children less than 16 years was 1.71 per million children.[19] There was a bimodal age distribution with incidence commoner in under fives and adolescents. Gender did not influence outcome. Complete recovery occurred in over 50% of children and fair/poor recovery occurred in over 25%. A better prognosis was associated with being under 10 years old; lesions of the lumbar spine; and preceding infection. A worse prognosis was associated with leg flaccidity; urinary incontinence; and rapid onset (peak dysfunction within 12 h). This study did not state how recovery was quantified, and excluded spinal cord compression and trauma[19]

A large all-cause series in young children under 5 showed that the most common aetiology was trauma from motor vehicle accidents occurring in 49% of cases ($n = 159$).[16] Complete injuries (ASIA A, see Methods) were associated with paraplegia rather than tetraplegia; motor vehicle accidents and violent aetiologies; and complications such as autonomic dysreflexia, severe scoliosis, hip dysplasia, pressure ulcers and bladder/bowel dysfunction. At follow up, community ambulators (24/159) were more likely to have had ASIA D lesions at presentation. ASIA A, B and C patients were more likely to use manual or power assisted wheelchairs. This was a single-centre study from a supra-regional specialist unit which may thus be a selected population.[16]

Data on causes and early predictors of outcome for acute paraplegia in children are limited. There is no uniform data collection system for this group of children in the UK. The objectives of this case series review were to describe the epidemiology of childhood-onset acute paraplegia and identify early prognostic factors (i.e. data available within the first few days of presentation that are associated with outcome). Such data may be of clinical value in counselling parents and young people in the very difficult days after onset of paraplegia.

2. Methodology

Attempts were made to ascertain all presentations of acute paraplegia in children within the former Northern region of England (Northumberland, Tyne & Wear, Durham, Tees-side and Northern Cumbria) during the period 1997 to March 2012. In the 2011 Census this area had a total population of 2,923,999 and an under 16 population of 601,263.[20] Cases were ascertained and cross-referenced from three sources: (i) the departmental database for all children seen in the single regional paediatric neurology service, (ii) children known to the single regional paediatric neuro-oncology service at the same hospital and (iii) ascertainment via the same hospital's neuroradiology database, identifying all children having MR imaging of the spine during this period. The regional adults' spinal injury unit is at a different hospital and does not generally accept paediatric patients. A small number of older adolescents were identified as having passed through this unit, however all had been identified from the other ascertainment sources.

The inclusion criterion was acute or sub-acute onset, partial or total, paraplegia in a child under 16 at presentation. As the aim of the study was to identify patients who had presented acutely with pathology to the spinal cord we excluded: confirmed cases of acute or chronic inflammatory demyelinating polyneuropathy (Guillain–Barré syndrome and CIDP); presumed or proven hereditary spastic paraparesis; and cerebral palsy and other developmental/congenital conditions including neural tube defects. We also excluded dissociative (functional) paralysis, peripheral neuromuscular disease,