RESULTS OF ANALYSIS

The average aluminium intake was 14 µg/day. Total intake of aluminium from intravenous feeding solutions was 645 µg. Additional aluminium may have been given with intravenous drugs and plasma, and with oral feeds.³

The aluminium content of the subject's grey matter was 40.1 µg/g wet weight of tissue; the mean (SD) value for 12 infants dying unexpectedly within the first year of life was 2.4 (1.6) µg/g wet weight of tissue. The fat content of the brain doubles during the first year of life; this would not however, account for the 20 fold difference in brain aluminium concentrations observed. In addition, aluminium accumulates specifically in cortical grey matter, the fat content of which is less likely to vary with age.

Discussion

The preterm infant studied here had a brain aluminium content of 40 µg/g wet weight of tissue. In 'dialysis dementia', corresponding cortical aluminium concentrations are reported to be 20–30 µg/g in adults,¹ and even lower in infants.^{2,4}

Toxicity is not proved by the finding of a high brain aluminium content. Nevertheless, with a history of unexplained convulsions and prolonged intravenous feeding, there is a strong possibility that high aluminium intake was at least a large contributory factor.

Parenterally administered aluminium is poorly excreted by preterm infants, who retain up to 80%

of an intravenous load.⁵ Calcium gluconate, potassium acid phosphate, and the trace element solution (Ped-EI) together accounted for 90% of the contamination. The substitution of calcium chloride for calcium gluconate reduces delivery of aluminium by 70% from an average intake of 30 µg/kg/day when on full intravenous feeding at 150 ml/kg/day to 8 µg/kg/day. The lower value of 14 µg/kg/day for the index case was due to partial enteral feeding.

The signs of aluminium toxicity, which include encephalopathy, anaemia, and poor bone mineralisation, are common complications of many diseases in sick premature babies. Nevertheless, we suggest that when conventional therapeutic manoeuvres fail, aluminium toxicity should be considered.

References

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Gowers' sign revisited

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SUMMARY We studied the patterns of standing in children with neuromuscular disorders and central hypotonia for Gowers' sign, and compared these two groups with healthy controls. More children with central hypotonia than controls rolled prone before standing at 36 months; at this age all children with neuromuscular disorders rolled prone. Neurological assessment is indicated in children continuing to roll prone before standing at 3 years.

In 1879 Gowers first described the pattern of standing in 21 boys with pseudohypertrophic muscular paralysis in a clinical lecture to the students of

University College.¹ He initially thought this pattern of standing was pathognomonic for children with this condition as it was present in all his ambulatory cases. It has subsequently been shown to be present in other children with proximal muscle weakness.

Dr Gowers' eloquent description of the pattern of standing, which now bears his name, emphasised two important features (figure): (i) the children adopting a prone position on all fours before attempting to stand and (ii) the children 'walking up their legs'. It is the second feature that is often quoted in textbooks and remembered by physicians as Gowers' sign.

The Muscular Dystrophy Group report that the

Figure Gowers' sign. Reproduced with kind permission of the publishers from WR Gowers. *Clinical lecture on pseudohypertrophic muscular paralysis. Lancet*, 1879.¹

mean age for diagnosis of boys with Duchenne muscular dystrophy is 5.2 years whereas parents first present their children to doctors at a mean age 2.7 years.

We believe the mean delay in diagnosis of 2.5 years is largely due to doctors failing to recognise the salient feature of Gowers' sign—that is, the child adopting a prone position before standing. It is only

when proximal leg weakness becomes most pronounced that children 'walk up their legs'.

The adoption of a prone position before standing can also be seen as a normal developmental phase in toddlers. This study defines the age at which children cease to turn prone while rising to a standing position and then examines the pattern of standing in children known to have neuromuscular disorders or central hypotonia.

Patients and methods

The study group consisted of 23 children aged 27–90 months (mean 58 months) with neuromuscular disorders and 26 children aged 16–69 months (mean 37 months) with central hypotonia. These children were consecutive new referrals seen by GBW at paediatric neurology and neuromuscular clinics at the Royal Manchester and Booth Hall Children's Hospitals.

The children with central hypotonia had no evidence of a neuromuscular disorder on clinical, neurophysiological, and in some cases, histopathological grounds. Controls consisted of 175 healthy children aged 13–60 months (mean 31.5 months) examined at district community child health centres.

The pattern of standing was defined by asking each child to lie supine and then to stand as quickly as they could on three separate occasions. Results were analysed according to whether the child adopted a prone position on two of those occasions at any time during standing. Frequencies between groups were compared using χ^2 analysis or Fisher's exact test where appropriate.

Results

Eighteen of 95 controls (19%) adopted a prone position as they stood at age 30 months or more; by 36 months only four of 63 (6.5%) rolled prone. Significantly more children with central hypotonia rolled prone, 15 of 20 (75%) at 30 months ($p < 0.001$) and eight of 13 (61%) at 36 months ($p < 0.001$). At

Table Pattern of standing in children ≥ 36 months. Results are No (%)

	Controls (n=59)	Neuromuscular disorder or central hypotonia (n=34)
Rolls prone:		
Yes	4 (7)	29 (85)
No	55 (93)	5 (15)

$\chi^2 = 66$, $p < 0.001$.

no age did the group with neuromuscular disorders stop rolling prone when attempting to stand.

When the combined groups with neuromuscular disorders and central hypotonia were compared with controls the age of 36 months was significantly discriminating in distinguishing normal children from those with underlying neurological abnormality (table).

Discussion

Our study emphasises the salient features of Gowers' sign and its importance in identifying children with neuromuscular disorders and central hypotonia. It is

easily performed and can readily be incorporated into screening examinations of children.

We suggest that if children continue to roll prone when attempting to stand at 30 months special attention should be paid to them. If they continue to roll prone at 3 years of age a full neurological assessment is warranted.

Reference

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