attempting to treat children without irradiation, but giving instead high doses of parenteral and intrathecal methotrexate to treat the occult disease of the central nervous system. Some recent studies have suggested that identical intellectual deficits may also be seen in these children suggesting either that the methotrexate causes similar problems to irradiation, or that intellectual damage might be caused by the disease itself.

There is no unanimity, however, in the reports of intellectual deficits in children with acute lymphoblastic leukaemia. Meadows *et al* were unable to show the same intellectual deficits in children treated with 18 Gy as they had described with 24 Gy.⁵ Eiser has recently reviewed the whole problem of cognitive deficits in children treated for leukaemia.⁶ Clearly they do occur in

many children and for some these are an educational and social handicap. Irrespective of the underlying cause of the problem, remedial help is warranted.

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Congenital neuroblastoma presenting with paraplegia

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Abstract

A girl presented with paraplegia at birth. A magnetic resonance scan showed an abdominal neuroblastoma with intraspinal extension. Treatment by chemotherapy did not produce any improvement in the neurological signs. Review of previously reported cases suggests the outlook for neurological recovery is poor regardless of treatment.

Case report

A newborn girl, delivered at 38 weeks' gestation by mid cavity forceps, was noted to have bilateral drop foot, a patulous anus with bulging of the perineum, and bilateral subluxing hips. In the hours after birth she dribbled urine

continuously. Plain spinal x ray films showed expansion of the spinal canal between T12 and L2. A magnetic resonance scan showed an extradural tumour extending from T12 to L3, continuous through the intervertebral foramina with a paraspinal mass on the left side (figure). Her homovanillic acid concentration was 38.8 $\mu\text{mol}/\text{mmol}$ creatinine (normal= <20) and hydroxymethoxymandellic acid 30.4 $\mu\text{mol}/\text{mmol}$ creatinine (normal= <9) supporting a diagnosis of neuroblastoma. A biopsy specimen was taken from the abdominal portion of the tumour at laparotomy. Histology showed this to be a poorly differentiated neuroblastoma. At surgery the liver appeared normal and there were no identifiable lymph nodes. The baby was treated with chemotherapy (vincristine 1.5 mg/m^2 , carboplatin 400 mg/m^2 , etoposide 150 mg/m^2 , and cyclophosphamide 600 mg/m^2).

This was well tolerated but there was no improvement in the neurological signs. Her unstable hips were managed in a Von-Rosen splint and her drop foot by ankle-foot orthoses.

Discussion

Congenital neuroblastoma presenting with paraplegia is rare. We have found another 15 cases reported in the literature (table).¹⁻³ In all but one case the tumour appeared to be localised at diagnosis, the exception having hepatic metastases. The treatment given was variable. The majority underwent decompressive laminectomy with, in most cases, postoperative radiotherapy or chemotherapy. Two were treated by chemotherapy alone and none by radiotherapy alone, and one by vitamin B-12 alone. Survival appears good with only two deaths; however, the neurological outcome was very poor.

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Magnetic resonance scan showing paraspinal tumour (open arrow) with intraspinal extension (black arrow).

Congenital neuroblastoma with paraplegia

Author*	Age at diagnosis	Treatment	Outcome	Neurological outcome	Follow up period (years)
Moschos and Anagnostakis (1975)	6 Days	Laminectomy, chemotherapy	Alive	Improved sphincter tone, paraplegic	1·9
Kenney <i>et al</i> (1982) ¹	5 Weeks	Chemotherapy	Died	No improvement	0·3
Bodian (1963)	Not known	Laminectomy, chemotherapy	Died	No improvement	0·3
	10 Months	Laminectomy, vitamin B-12	Alive	No improvement	2·3
	3 Weeks	Biopsy, vitamin B-12	Alive	No improvement	6·6
	6 Days	Laminectomy, radiotherapy, vitamin B-12	Alive	No improvement	1·8
Katcher (1952)	3 Weeks	Laminectomy, radiotherapy, chemotherapy	Alive	Improved sphincter tone, persisting limb weakness	0·3
Hrabovsky and Jones (1979)	2 Weeks	Laminectomy, chemotherapy	Alive	No improvement	1·0
Elefant <i>et al</i> (1958)	1 Month	Laminectomy, radiotherapy	Alive	Improved sphincter tone, persisting limb weakness	1·7
Punt <i>et al</i> (1980) ²	3 Days	Laminectomy, radiotherapy	Alive	No improvement	14·0
	1 Day	Laminectomy	Alive	No improvement	11·0
	4 Months	Laminectomy, radiotherapy	Alive	No improvement	7·0
	2 Weeks	Radiotherapy, chemotherapy	Alive	No improvement	4·0
Haden and Keats (1983) ³	1 Month	Laminectomy, radiotherapy	Alive	Residual weakness of left leg	32·0
Rothner (1971)	2 Days	Laminectomy, radiotherapy	Alive	No improvement	0·8
Munro <i>et al</i> (1991)	3 Days	Chemotherapy	Alive	No improvement	0·8

*The full references for those publications not included in the list of references at the end of the paper can be obtained from Kenney *et al*,¹ Punt *et al*,² and Haden and Keats.³

Twelve patients showed no improvement in their neurological signs after treatment and four showed some improvement but were still left with significant neurological deficits. The method of treatment or the time from birth to treatment did not seem to be different between those showing some recovery and those with none. By comparison series of older children with cord compression from neuroblastoma show poorer survival, particularly in those over 1 year of age at presentation, but a much better neurological outcome. Series of patients treated both surgically^{2 4 5} and by chemotherapy alone⁶ have shown neurological recovery in 40–80% of cases. Surgery is not without its complications: significant spinal deformity is reported as affecting up to 60% of survivors after laminectomy.^{4 5} Given this high morbidity and the seemingly poor prognosis for neurological recovery we

would question the usefulness of laminectomy in congenital neuroblastoma with paraplegia.

We would postulate that in this group cord compression must occur antenatally and that by the time of birth, irreversible damage has already occurred.

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Omenn’s disease

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Abstract
The importance of accurate pathological diagnosis is emphasised in the case of a newborn infant who presented with alopecia, a generalised erythrodermatous skin eruption, and hepatosplenomegaly. She subsequently developed generalised lymphadenopathy and recurrent septicaemia and died aged 2 months. The histological findings of widespread lymphocytic, histiocytic, and eosinophilic tissue infiltration, associated with thymic hypoplasia, were consistent with autosomal recessive Omenn’s disease.

Omenn’s disease is a rare autosomal recessive reticuloendotheliosis of infancy with similarities

to many other reticuloses but with several distinguishing features of genetic and therapeutic importance. We are unaware of any previously reported cases in the United Kingdom.

Case report
The index case was a girl who was the first child of non-consanguineous parents. The pregnancy and delivery were uncomplicated and her birth-weight was 2830 g. Hair was seen in the liquor and remaining scanty scalp hair was wiped off when she was dried. There was no other bodily hair but a generalised dry scaly rash was present with flexural cracking and extensive serous exudation. Liver and spleen were both palpable 2 cm below the costal margins and the abdomen

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