PERMANENT FLACCID PARAPLEGIA IN CHILDREN WITH THORACIC SPINAL CORD INJURY*

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Abstract: Among 12 children with complete thoracic spinal cord injury four developed permanent flaccid paraplegia with areflexia and an autonomous bladder. Clinical, neurophysiological and myelographic findings favour an extensive lower motor neurone lesion (myelomalacia).

Key words: Spinal cord injury; Spinal cord infarction; Flaccid paraplegia; Myelography; Nerve conduction velocity.

Introduction

In the acute phase of traumatic spinal cord injury flaccid paraplegia and areflexia are constant features. Once the stage of spinal shock subsides there is reflex return and some degree of hyperreflexia and spasticity evolves. This applies to trauma to the thoracic cord while it is well recognised that lesions of the lumbosacral cord and cauda equina commonly result in flaccid paraplegia. Failure of reflex return in thoracic lesions is unusual and destruction of grey matter in the vertical plane seems to be prerequisite (Guttmann, 1973).

When we reviewed a series of children who sustained spinal cord injuries we were surprised to find four cases with permanent flaccid paraplegia following thoracic cord trauma. These cases are the topic of this report.

Material and Methods

Patients

From January 1960 to March 1979 25 children with spinal cord trauma were admitted to our hospital. Details with particular reference to long-term rehabilitation have been reported elsewhere (Bucher *et al.*, 1980). The age at injury was below 12 years, apart from two patients (13 years and 16 years respectively). The transverse lesions were incomplete in nine and complete in 14 children, while two cases were lost to follow-up. There were 12 complete lesions with a neurological level above T12 segment. In the period under review 10 newborns with birth injury to the spinal cord were seen. They have been described separately (Bucher *et al.*, 1979) and will not be discussed further.

Nerve conduction velocity (NCV)

Using DISA equipment NCV measurements were obtained in the median nerve. Measurements in the lower limbs were attempted in both tibial and peroneal nerves using surface stimulating electrodes and needle recording electrodes. Registration of H-reflex was tried by stimulation of the tibial nerve in the poplitea and recording over the m. triceps surae.

* Dedicated to Prof. A. Prader on the occasion of his 60th birthday.

Myelography

Lumbar myelography, using metrizamide (Amipaque), was performed in two children (cases 1 and 2) without local or general anaesthesia. There were no adverse side-effects. The air myelogram shown for comparison in Fig. 3 was performed under general anaesthesia.

Description of Cases

Table I summarises some relevant data of our four cases with permanent flaccid paraplegia. Before further details are added below, a description of findings common to all patients follows.

TABLE I

Case/Sex	Age (years)		Neurol. segment of cord lesion	Ambulation	Last IVP	Other remarks
	At injury	At present				
1 f	1½	5	T7	Rollator	1978 (July) normal	Neurofibromatosis Glioma optic chiasm Myelography—Fig. 1
2 m	9¾	14	T7	Swing-through gait	1979 (July) normal	Myelography—Fig. 2
3 m	5½	18½	T9	Swing-through gait	1976 normal	Marked scoliosis
4 m	11¼	25	T8	Drives car Wheel-chair Drives car	1979 (July) Mild hydro- nephrosis	Marked scoliosis Diabetes mellitus

General remarks

All cases sustained comparatively minor trauma to the spine. While the accident in case 1 led immediately to a complete paraplegia, the neurological deficit resulting in complete transverse lesions evolved within half an hour in cases 2 and 4, and was realised only some hours later in case 3. Radiographs revealed no bone injuries. The transverse lesions remained complete and there was no sacral sparing of sensation. The lower limbs remained completely flaccid with muscular wasting, absence of tendon reflexes and complete sensory loss. Abdominal, anal (and in males cremasteric) reflexes were abolished. Plantar stimulation brought no response. There were no foot deformities but cases 3 and 4 have considerable scoliosis. There were no para-articular ossifications.

Case 1. This 1½ year-old girl was thrown out of her push-chair which was hit by a car. She had immediate complete paraplegia with a sensory level at the lower costal margin. Myelography was normal and CSF was clear. Her neurological condition did not change subsequently and she remained flaccid and areflexic in her legs. So far she has had four fractures of her lower legs due to osteoporosis. She had recurrent urinary tract infections but intravenous pyelography was completely normal (August 1978). Bladder rehabilitation with regular suprapubic tapping was not successful and her bladder has to be emptied by gentle manual pressure.

Already on her first admission multiple large café-au-lait spots were recognised and neurofibromatosis was considered. At the age of 4½ years strabismus was noted. There was bilateral optic atrophy. An optic chiasm glioma was suspected and confirmed by computed tomography. Myelography was performed at 4¾ years (Fig. 1).

FIG. 1

Case 1. Complete lesion corresponding to T7 segment. Metrizamide myelography, A.P. view (Fig. 1a): Irregular contour of spinal cord in mid-thoracic region. Cord not clearly visualised below T8 vertebra. Lateral view (Fig. 1b): The spinal cord tapers to a point at T8/9 vertebral level.

Case 2. This 9 $\frac{3}{4}$ -year-old boy fell from a van on his buttocks. He experienced marked backache but he managed to get up and walk a few steps before he collapsed because of weakness of his legs. Details of his initial neurological examination are not known but he was subsequently found to have persistent complete flaccid paraplegia corresponding T7 segment. Rehabilitation was successful in so far as he reached complete independence. He is able to walk long distances by swing-through gait. He had no ulcers, no fractures, no urinary tract infections and he has only mild scoliosis. However he did not develop an

FIG. 2

Case 2. Complete lesion corresponding to T7 segment. Metrizamide myelography, A.P. view: Cone-shaped spinal cord at T8 vertebral level, not recognised below this point.

automatic bladder and he only voids following suprapubic tapping by gentle manual pressure. He had a normal intravenous pyelogram in July 1979. Myelography was performed at 14½ years (Fig. 2).

Case 3. This patient was said to have fallen out of his bed at night when he was 5½ years old. He complained immediately of pain and weakness in his lower limbs. The following morning he woke up paraplegic and incontinent. CSF examination revealed clear fluid. He had a permanent complete cord lesion with flaccidity and areflexia corresponding to a level of T9. He gained complete independence, and walks up to 10 km by a swing-through gait; he has no wheel-chair and he drives a car. There have been several skin ulcers and three fractures due to falls. He has not had any urinary tract infection, but he failed to develop an automatic bladder and voids with abdominal pressure. The last IVP (1976) was normal. He refused a repeat IVP and urodynamic studies.

Case 4. This patient has had diabetes mellitus since he was aged 3 years. At the age of 11, during gymnastics he fell onto his back from a height of about 1½ metres, had intense pain and experienced some weakness of his lower limbs. Within half an hour he had complete paraplegia. On admission the CSF was normal as was myelography. A repeated myelogram 6 weeks later showed a questionable arrest of the contrast at T3/4 vertebral level. Laminectomy was performed (in 1966), but no abnormality was seen. A complete cord lesion with a level corresponding T8 segment persisted. This patient reached independence but had recurrent urinary tract infections despite good control of his diabetes mellitus. Pyelography showed moderate hydronephrosis. He has an autonomous bladder with markedly increased capacity, uses a urinal and voids with manual pressure. There have been no fractures, but recurrent skin ulcers pose great problems.

Results of NCV

NCV was within normal limits in the upper limbs in all four cases. However H-reflexes and motor NCV were not measurable in the lower limbs. On the other

hand NCV were easily recordable and found to be normal in seven children with spastic traumatic paraplegia.

Results of Myelography

Representative aspects and corresponding line drawings of myelographs of cases 1 and 2 are shown in Figs 1 and 2 (for description see legends). CSF analysis was normal in both cases.

For comparison, in Fig. 3 an air myelograph is shown of a four-year-old girl with a diagnosis of transverse myelitis one year previously. She had persistent flaccid paraplegia, areflexia of her lower limbs and urinary incontinence due to a

FIG. 3

Air myelography, lateral view. Four-year-old girl with complete lesion corresponding to T4/5 segment, one year after transverse myelitis. The spinal cord tapers off below T4 vertebra to become string-like at and below T6 vertebra.

complete transverse lesion at T4/5 segment. Follow-up studies show that her condition has been unchanged in the past 3 years. She has an autonomous bladder.

Discussion

In the four cases described an additional lower motor neurone lesion resulting in flaccidity and areflexia had to be assumed. This was supported by neurophysiological investigations (i.e. absence of recordable NCV) and in two patients further substantiated by radiological evidence of myelomalacia. In view of the assumption of a lower motor neurone lesion at spinal (anterior horn) level it was interesting to realise that these patients clinically have an autonomous bladder, although unfortunately urodynamic and sphincter electromyographic studies are not available. Despite intensive training these patients did not succeed in emptying their bladder by suprapubic tapping but they void at regular intervals by manual pressure, partly assisted by abdominal pressure. This can be explained by destruction of the spinal bladder centre. We speculate that permanent flaccidity following thoracic cord trauma as a rule coincides with an autonomous bladder, although this

is not explicitly clear from the literature. From the urological point of view this may be disadvantageous but, on the other hand, it is of benefit to these patients not to be subject to flexor-adductor spasms and clonus of the lower limbs. In our limited experience flaccidity certainly does not impede rehabilitation for ambulation.

Regarding pathogenesis, it is worth emphasising that cases 2-4 had sustained only comparatively minor trauma to the spine and that a few minutes to about half an hour elapsed before paraplegia was complete. This points to vascular factors being inflicted although the exact mechanism and the pattern of the assumed infarction is open to speculation. In view of the complete transverse lesions it is very unlikely that occlusion of the anterior spinal artery or the arteria radicularis magna of Adamkiewicz had occurred. Haemorrhagic infarction from a pre-existing spinal vascular malformation seems also most improbable as the CSF was clear. General severe hypoxia or drop in perfusion pressure which can be causative in spinal stroke (Silver and Buxton, 1966) did not occur in our patients.

In two large adult series (Guttmann & Frankel, 1966; Guttmann & Walsh, 1971) the frequency of complete flaccidity in thoracic cord trauma can be calculated to be 12 per cent. Other reports mentioning permanent flaccidity do not state the neurological level nor do they correlate the clinical observations with neurophysiological, urological or even neuroradiological investigations (Rivera-Dominguez *et al.*, 1978-79; Girard *et al.*, 1978-79). Judging from the scant literature, flaccid paralysis in children with thoracic cord injury is very rare or has not created great interest.

In larger paediatric series this phenomenon is not even mentioned (Meinecke, 1972; Hachen, 1977-78). Burke (1971) described two young children with permanent flaccid paralysis following cord injury at T3 and T4 segment. Ahmann *et al.* (1975) reported two patients in whom spinal cord infarction due to minor trauma was confirmed by pathological study. One case was flaccid in the upper limbs only, while the second case was tetraplegic and areflexic.

Permanent flaccidity and areflexia has occasionally been observed following birth trauma to the spinal cord (Allen, 1970; Fontan *et al.*, 1964; Jellinger & Schwingshackl, 1973). In our personal series of 10 cases (Bucher *et al.*) one boy with a level at T4 segment remained flaccid and areflexic and NCV was not measurable in the lower limbs. Some cases with transverse myelitis also result in permanent flaccidity (Ropper & Poskanzer, 1978). Fig. 3 shows the myelogram of a personally examined child diagnosed as having had transverse myelitis one year previously. Extensive spinal cord destruction (myelomalacia), presumably on a vascular basis, seems to be responsible for her permanent flaccidity.

SUMMARY

From January 1960 to March 1979 25 children with spinal cord injury were admitted to our hospital (10 newborns with birth injury to the spinal cord were excluded). Among 12 patients with complete thoracic lesions four remained permanently flaccid. These four cases who had sustained relatively minor trauma showed marked muscular atrophy of the lower limbs, areflexia, absence of anal and cremasteric reflexes, no response to plantar stimulation and no foot deformities. In contrast to children with spastic traumatic paraplegia, motor nerve conduction velocities and H-reflexes were not measurable in these flaccid patients. Myelography was performed in two, this showing myelomalacia below the level of injury. All four patients had clinically an autonomous bladder and voided by gentle

manual pressure. The clinical, neurophysiological and radiological findings are consistent with a lower motor neurone lesion below the level of cord injury, resulting presumably from an extensive longitudinal cord lesion on a vascular basis. Judging from Guttman's experience, flaccid paraplegia occurs in about 12 per cent of adults with complete thoracic cord lesions. The literature is too scant to give an estimate of this complication in children with traumatic paraplegia.

RÉSUMÉ

Vingt-cinq enfants avec une paraplégie traumatique furent hospitalisés dans notre clinique entre le début de l'année 1960 et mars 1979 (non compris 10 nouveau-nés avec une lésion de la moelle épinière due à un traumatisme lors de l'accouchement). Quatre patients parmi les 12 qui avaient une lésion transverse complète de la moelle épinière au niveau thoracique restèrent complètement paralysés avec une paralysie flasque. Ces quatre patients, qui avaient souffert par ailleurs d'un traumatisme relativement insignifiant, présentèrent les signes suivants: atrophie musculaire nette des jambes, aréflexie tendineuse, inhibition du réflexe anal et crémasterien. Aucun ne développa une déformation des pieds. Le réflexe H et la vitesse de conduction nerveuse mesurés aux jambes n'étaient pas enregistrables contrairement aux enfants atteints d'une paralysie spastique. Une myélographie fut faite chez deux patients et montra une myélomalacie en dessous du niveau de la lésion transverse. Les quatre patients présentèrent cliniquement une vessie autonome qui pouvait être vidée manuellement. Les examens cliniques, neurophysiologiques et radiologiques parlent en faveur d'une dégénérescence du deuxième motoneurone situé en dessous de la lésion transverse due probablement à des troubles vasculaires. Dans des séries étendues de Guttman se référant aux adultes 12 pourcents des lésions transverses complètes de la moelle épinière thoracique évoluent vers une paraplégie flasque. Il n'existe pas dans la littérature des résultats analogues chez l'enfant.

ZUSAMMENFASSUNG

Von Januar 1960 bis März 1979 wurden in unserer Klinik 25 Kinder mit traumatischer Paraplegie hospitalisiert (10 Neugeborene mit geburts-traumatischen Rückenmarksverletzungen nicht eingeschlossen). Von 12 Patienten mit vollständiger thorakaler Läsion blieben 4 völlig schlaff. Klinisch waren diese Fälle, die ein relativ geringfügiges Trauma erlitten hatten, gekennzeichnet durch deutliche Muskelatrophie an den Beinen, Areflexie sowie Fehlen von Anal- und Cremasterreflex. Es bestanden keine Fußdeformitäten. Im Gegensatz zu Kindern mit spastischen traumatischen Paraplegien konnten bei den schlaffen Patienten an den Beinen keine Nervenleitgeschwindigkeiten und kein H-Reflex registriert werden. Die bei 2 Kindern durchgeführte Myelographie ergab eine Myelomalazie unterhalb des Niveaus der Querschnittsläsion. Klinisch besteht bei allen 4 Patienten eine autonome Blase, die durch vorsichtige manuelle Expression entleert wird. Klinische, neurophysiologische und neuroradiologische Befunde sprechen für einen Ausfall des unteren Motoneurons unterhalb der Querschnittsläsion, vermutlich auf vaskulärer Grundlage. In Guttman's umfangreichen Serien sind 12 Prozent der vollständigen thorakalen Querschnittslähmungen Erwachsener in eine schlaffe Paraplegie übergegangen. Entsprechende Zahlen sind für das Kindesalter nicht verfügbar.

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