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Split cervical spinal cord with Klippel–Feil syndrome: seven cases

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Summary

We report seven cases of rare high cervical split spinal cord associated with extensive vertebral fusions (Klippel–Feil anomaly). In light of previous embryological theories and recent research findings we attempt to explain the origin of split cord and vertebral fusions. Two distinctly separate mechanisms are suggested for the development of split cords observed in our cases: a midline lesion bisecting the neuroepithelium and the notochordal plate could be responsible for complete splitting of the cervical cord with anterior bony defect while a localized disturbance of cervical neural tube closure would account for cases with partial dorsal splitting of the cord with posterior vertebral defect. Vertebral fusion anomalies are likely to be associated with disturbance of Pax-1 *gene expression in the developing vertebral column. We confirm with our cases the frequent association of failure of normal segmentation and split cord in the cervical region. Clinically, only three patients had neurological deficit which was mild and has remained stable, and they had no radiological evidence of tethering; the minimal disproportionate growth of the cord and spine and the rarity of a bony spur in the cervical region are the likely reasons. A conservative policy was therefore pursued in these cases with careful long-term follow-up.*

Keywords: congenital malformations; vertebral fusion; diastematomyelia; embryogenesis; *Pax* genes

Introduction

Diastematomyelia describes a rare double cord malformation which is usually diagnosed in childhood, occurs mainly in the lower thoracic and lumbar regions and is more common in females. The spinal cord is either completely or partially split over one or more segments, with or without a fibrous or osseocartilaginous septum (Herren and Edwards, 1940; Lichtenstein, 1940; Bligh, 1961; James and Lassman, 1972; Hilal *et al.*, 1974; Winter *et al.*, 1974; Naidich *et al.*, 1983; French, 1990; Pfeifer, 1991). Cervical examples are rare; so far only ~25 cases have been reported (James and Lassman, 1972; Giordano and Davidovits, 1982; Beyerl *et al.*, 1985; Okada *et al.*, 1986; Simpson and Rose, 1987; Wolf *et al.*, 1987; Ohwada *et al.*, 1989; Pfeifer, 1991; Rawanduzy and Murali, 1991; Pang, 1992; Ulmer *et al.*, 1993). Most of these were operated on and a bony spur was an unusual finding. Several of the reported cases had the appearance of limited myeloschisis of the cervical cord which is also quite rare (Avery and Rentfro, 1936; Gunderson and Solitare, 1968; Nagib *et al.*, 1987).

Here, we present seven cases of high cervical split cord associated with extensive vertebral fusions (Klippel–Feil anomaly; Klippel and Feil, 1912). The rarity and diversity of these cervical lesions and their relatively frequent association with failures of segmentation justify an attempt to integrate previous embryological theories with recent research findings on the role of gene expression in the embryonic development of the cervical spinal cord and vertebrae.

Observations

Case 1

This 54-year-old lady had a congenital neck deformity with the classic clinical triad of Klippel–Feil syndrome (short neck, low posterior hairline and restricted range of motion of the neck). She had been well for 49 years when she started to have increasing neck pain radiating down between her shoulder blades. Later she noticed paraesthesia in the right

arm and a weakening grip in the right hand. She also had difficulty in climbing stairs, as her legs felt weak. Pain typical of occipital neuralgia was felt on the left side.

Examination

Neurological examination revealed bilateral 6th nerve palsies which had been present since childhood, bilateral mild sensorineural hearing loss, and weakness of the right triceps and the intrinsic muscles of both hands. She also had a grade 4/5 spastic paraparesis of the lower limbs and diminished sensation of pinprick and light touch between C6 and T1 in the right arm. Her tendon reflexes were brisk with down-going plantars.

Radiological findings

The CT-myelography showed diastematomyelia of the cervical spinal cord without any bony or cartilaginous spur extending downwards from the medulla to C3 (Fig. 1A). The dysplastic and narrow vertebral bodies had a midline sagittal cleft (Fig. 1B). The foramen magnum and the spinal canal were wide and there were no signs of compression or tethering of the cord. The anterior arch of C1 was bifid and the odontoid process relatively large. Roentgenograms revealed extensive fusion between C2 and C7 vertebrae and the posterior arch of C1 was fused to the occiput (Fig. 1C). The clivus was short and the large odontoid appeared to articulate with it. No sign of instability could be detected.

The patient has been neurologically stable for 4 years and as her functional deficit remains mild no operation has been performed.

Case 2

This 11-year-old girl was born with a short and deformed neck and, due to respiratory insufficiency, needed ventilation 5 h after birth. A large cystic mass in the posterior mediastinum was detected by CT scan and was removed at 2.5 months. It was adherent to the prevertebral fascia and histologically it proved to be a neurenteric cyst. Her respiratory problems rapidly resolved postoperatively and she underwent further investigations for her neck deformity at 5 months.

Examination

Physical examination showed a very short neck with limited movements. She had no neurological deficit.

Radiological findings

The CT-myelography showed a very wide cervical spinal canal with a split cord between C1 and C7 (Fig. 2A). A bilobular intradural cystic mass extended between the hemicords, the anterior lobe of which seemed to merge into the body of one of the lower cervical vertebrae. Roentgenograms confirmed the anterior round defect in this area (the presumed site of an intra-extraspinal communication of the neurenteric cysts). Extensive fusion of the dysplastic cervical and upper thoracic vertebral bodies and laminae with a widened interpedicular distance were also obvious (Fig. 2B and C).

The patient has remained neurologically asymptomatic and no surgery on the cervical spine has been performed.

Case 3

This 13-year-old girl was born with a large occipital meningocele and a small epidermal cyst which were removed shortly after birth. A few days later a ventriculo-peritoneal shunt was inserted. At 2.5 years of age she started to display an intermittent head tilt but she remained otherwise asymptomatic until aged 12 years when she developed a progressive head tilt to the left associated with neck pain, both of which were thought to be partly due to fibrosis around the shunt tube (which passed down on the left side of the neck).

Examination

Physical examination confirmed the slight head tilt to the left with limited rotation to the right and slight flattening of the left side of her face. The neurological examination revealed no deficit.

Radiological findings

The cervical spine roentgenogram revealed a cervical scoliosis due to complex cross-fusions of multiple hemivertebrae between C2 and C7 on both sides. A midline cleft-like ossification defect was also visible in the middle cervical region, approximately at the level of C4–C5 vertebral segments (Fig. 3A). The MRI scan of this region demonstrated partial posterior splitting of the upper cervical cord without signs of any bony or cartilaginous spur (Fig. 3B and C). There were no signs of instability, cord tethering or compression.

The patient has undergone a re-routing of her distal shunt tube and this has improved her local pains, although her head tilt remains the same.

Case 4

This 24-year-old lady was born with Klippel–Feil syndrome associated with Sprengel's deformity. She was reported to have been asymptomatic as a child, but at the age of 12 years she stopped growing and at 20 years she was investigated for her various problems.

Examination

Physical examination revealed short stature, a short, webbed neck with low hairline and practically no cervical movement.

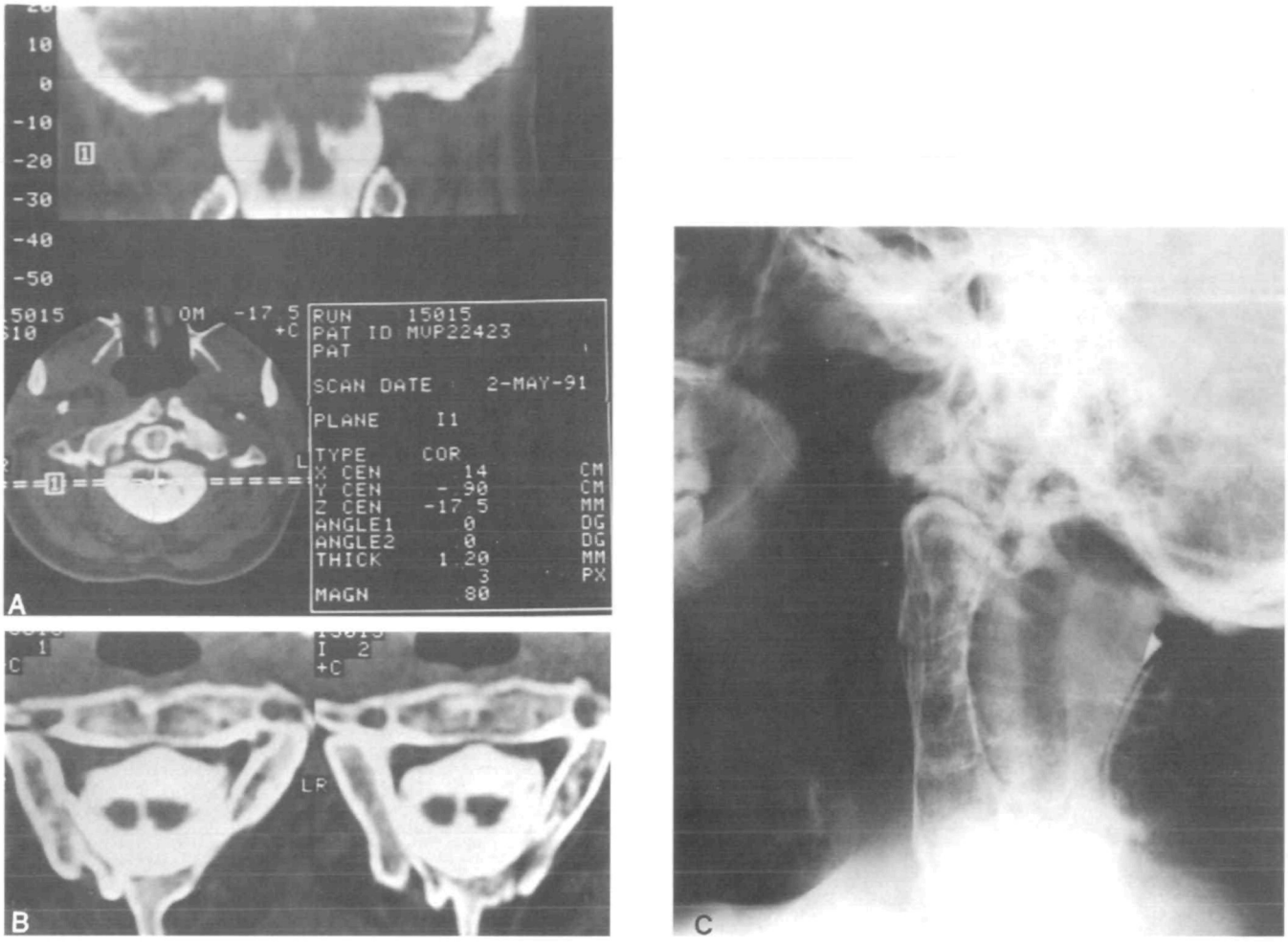

Fig. 1 Case 1. (**A**) Coronal reformatted CT-myelogram showing diastematomyelia between the medulla and C3. Note the anterior spina bifida of C1 and the large odontoid. (**B**) Axial CT-myelograms demonstrating dysplastic narrow vertebral bodies, with a thin zone of sclerosis in the sagittal plane indicative of an earlier midline defect resulting in separate ossification of right and left halves of each vertebral body and a wide spinal canal. A complete split of the cervical cord is evident in line with the defect of the vertebral bodies. (**C**) Lateral cervical spine roentgenogram (with intrathecal contrast material) showing extensive fusion between C2 and C7. The large odontoid appears to articulate directly with the short clivus, and there is a posterior assimilation of C1 to occiput.

She also had maxillary hypoplasia, a high arched palate, bifid uvula and submandibular hirsutism. The neurological examination showed strabismus, mirror movements, bilateral conductive hearing loss and minimal gait ataxia.

Radiological findings

The cervical spine roentgenograms demonstrated total fusion of hypoplastic cervical vertebrae with malformed and fused posterior elements. There was also a very wide foramen magnum and an extremely small posterior fossa (Fig. 4A). The MRI scan demonstrated splitting of the lower medulla and upper cervical cord without an osseocartilaginous spur or evidence of either compression or tethering. There was spina bifida and a meningocele in the upper cervical spine (Fig. 4B and C).

The patient has remained neurologically unchanged over the last 4 years and has been seen regularly in follow-up clinic.

Case 5

This 10-year-old girl was born with multiple congenital abnormalities including one lung, one kidney and heart valve insufficiencies for which she underwent cardiac surgery at the age of 6 months. She also had an occipital meningocele excised shortly after birth. She remained asymptomatic until the age of 7 years when she gradually developed neck pain.

Examination

Physical examination showed a slight head tilt to the left. Cervical flexion and extension were restricted to about 50% of normal. She had minimal rotation of the head. No neurological deficit could be detected.

Radiological findings

An MRI of the cervical spine revealed a partial dorsal wedge-shaped splitting of the spinal cord at the level of

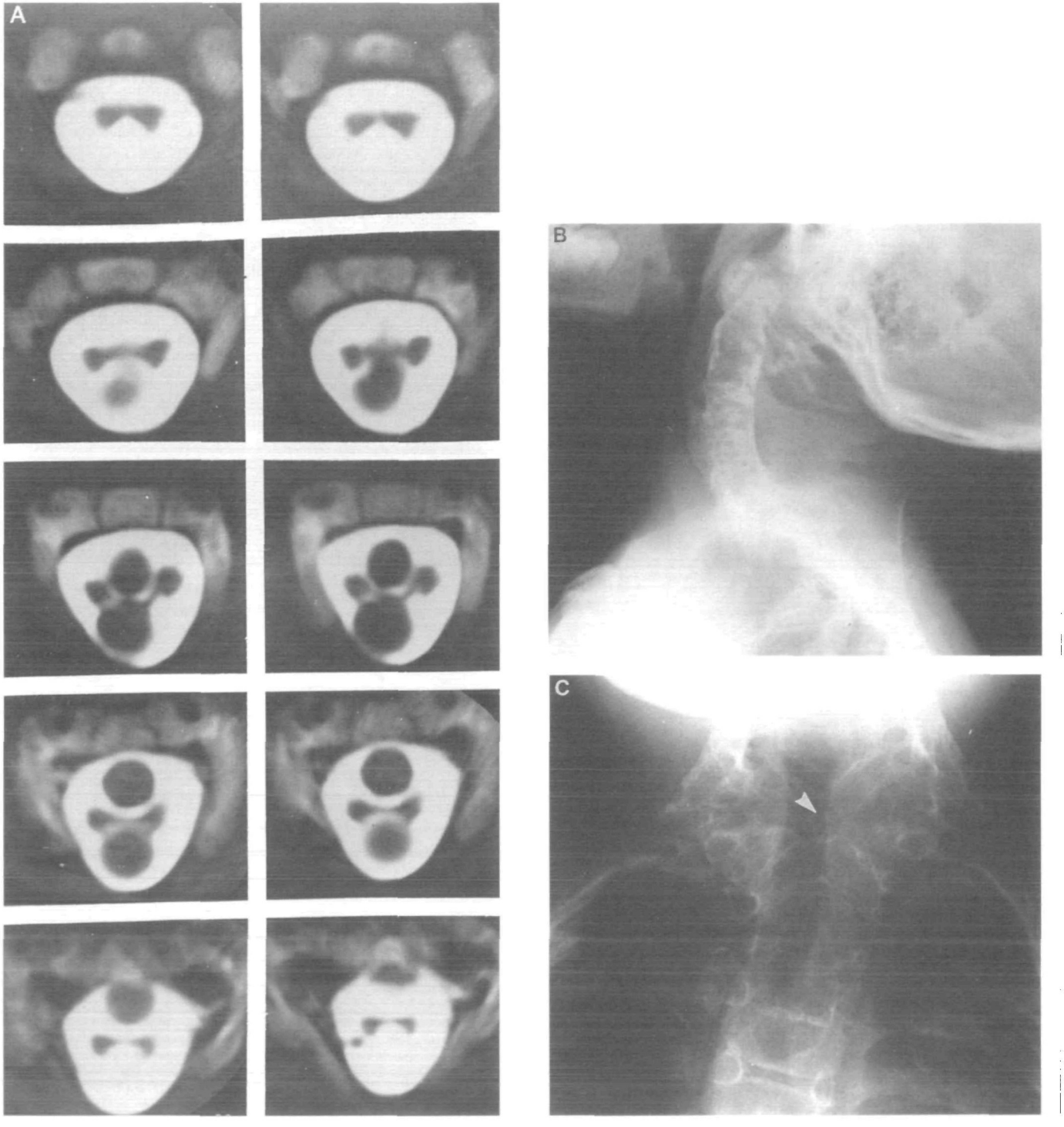

Fig. 2 Case 2. (**A**) Series of axial CT-myelograms between the foramen magnum and C7 at age 5 months showing a cervical cord widely split by the bilobular sagittally oriented cyst that lies in a wide dural sac. The anterior lobe of the intradural cyst appears to traverse the body of C7. (**B**) Lateral and (**C**) anteroposterior roentgenograms at age 10 years showing extensive fusions of the cervical and upper thoracic vertebrae, widened interpedicular distance and the round-shaped defect (arrowhead) marking the site of communication between the intraspinal and the previously excised posterior mediastinal neurenteric cysts.

C1–C3 without bony or cartilaginous spur (Fig. 5A). The foramen magnum was wide with minimally descended tonsils and an ununited odontoid peg (os odontoideum). There was no sign of compression or tethering of the cord (Fig. 5B). Flexion and extension cervical radiographs demonstrated extensive cervical fusion with C1/2 instability. The posterior elements were absent from most of the cervical vertebral bodies (Fig. 5C and D).

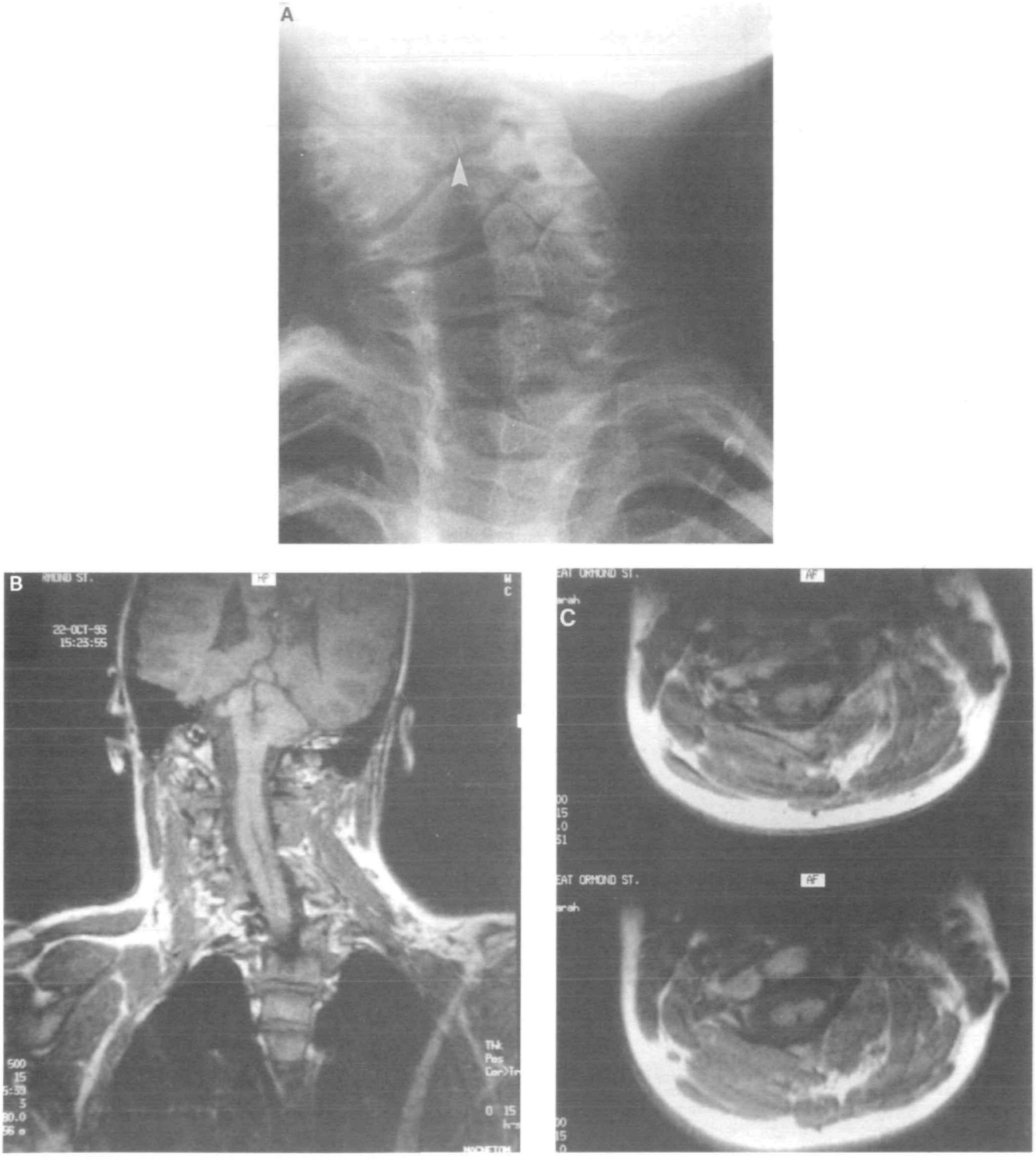

Fig. 3 Case 3. (**A**) Anteroposterior roentgenogram showing cervical scoliosis due to asymmetrically fused hemivertebrae on both sides. Note the sagittal cleft (arrowhead) which suggests, together with hemivertebrae and absent posterior bony defect, that this case belongs to the 'group with complete splitting of the cord'. (**B**) Coronal and (**C**) axial cervical MRI, T_1-weighted image exhibiting the posterior splitting of the cord.

The patient has been neurologically asymptomatic and a posterior occipitocervical fusion (occiput to C3) with rib graft was performed followed by application of halo orthosis.

Case 6

This 36-year-old lady has had the classic triad of Klippel–Feil syndrome since birth. She presented with a 2-year-

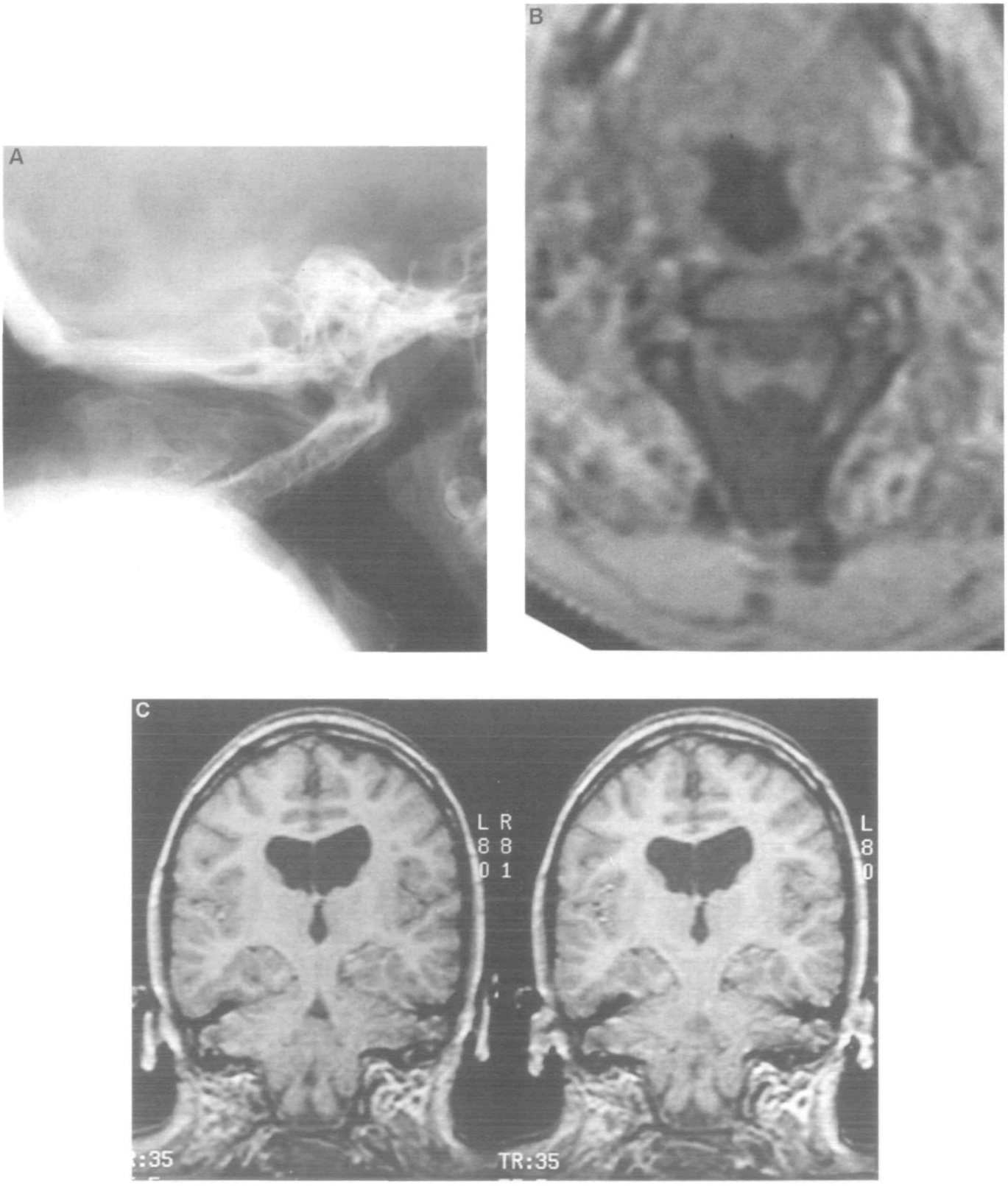

Fig. 4 Case 4. (**A**) Lateral cervical roentgenogram showing total fusion of hypoplastic vertebral bodies and fusion of malformed posterior elements. (**B**) Axial and (**C**) coronal MRI (T_1-weighted image) with dorsally split lower medulla and upper cervical cord. Note the wide spina bifida and the large dural sac on Fig. 4B.

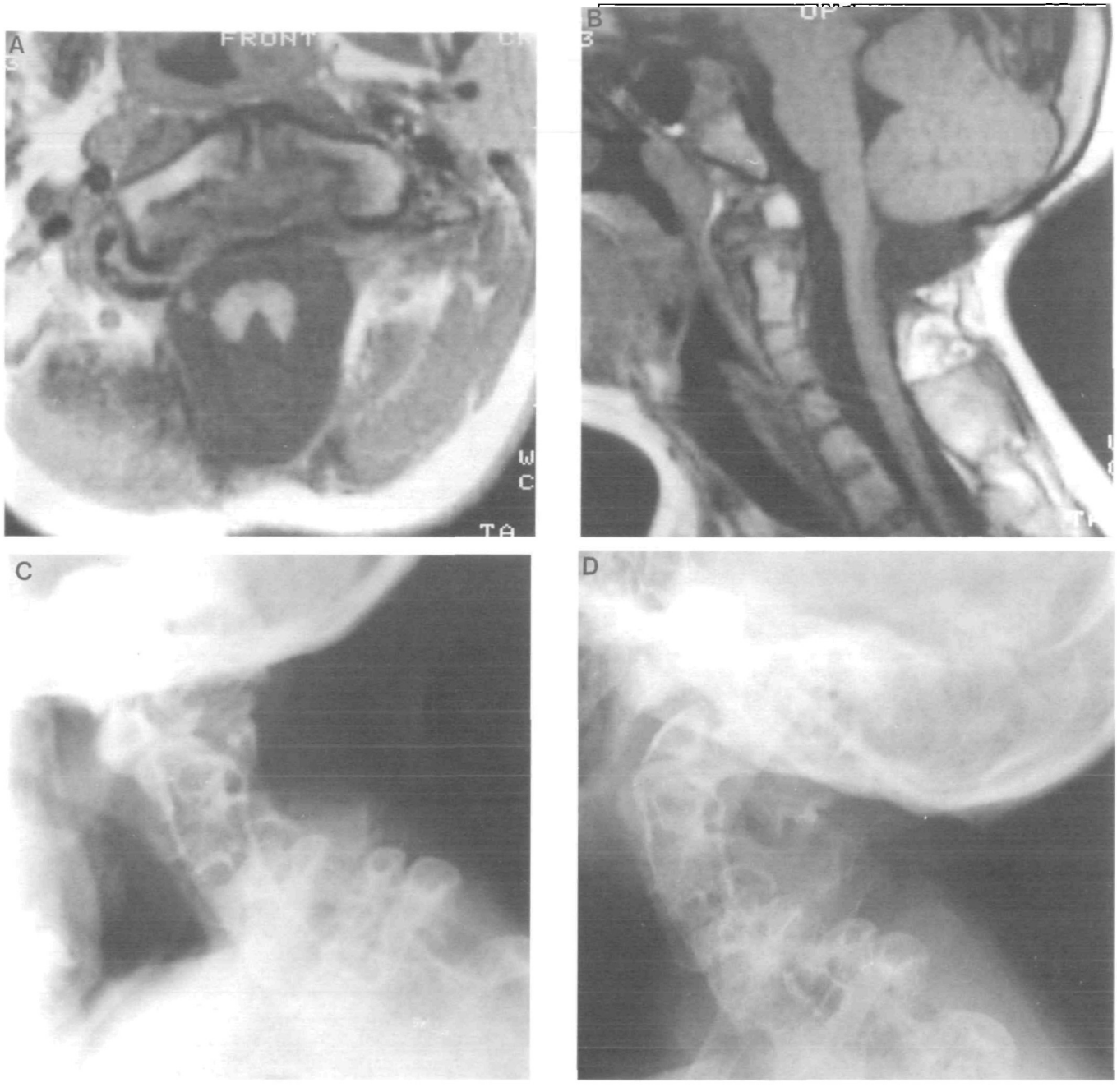

Fig. 5 Case 5. (**A**) Axial MRI (T1 weighted image) showing partial dorsal wedge-shaped splitting of the upper cervical cord. (**B**) sagittal MRI (T_1-weighted image) showing wide canal, vertebral fusion and os odontoideum. Extensive fusion and instability at C1/2 is demonstrated on (**C**) flexion and (**D**) extension lateral roentgenograms.

history of left occipital headache, dizzy spells and some episodes of blurred vision. Her personality also changed due to her pain; she became irritable. She had no other obvious associated anomaly.

Examination

Physical examination revealed a short neck with no movements and low hairline. She had short stature. Neurological examination showed a left temporal field defect but no deafness, nystagmus or other abnormality of the cranial nerves. There was marked tenderness over the left occipital nerve. She had a grade 4/5 spastic quadriparesis without any obvious sensory or sphincter abnormality.

Radiological findings

The cervical spine roentgenograms and MRI scan demonstrated fusion of the cervical vertebrae, a very wide foramen magnum and upper cervical spinal canal with small posterior fossa and without any sign of neural compression or tethering (Fig. 6A and B). A partial,

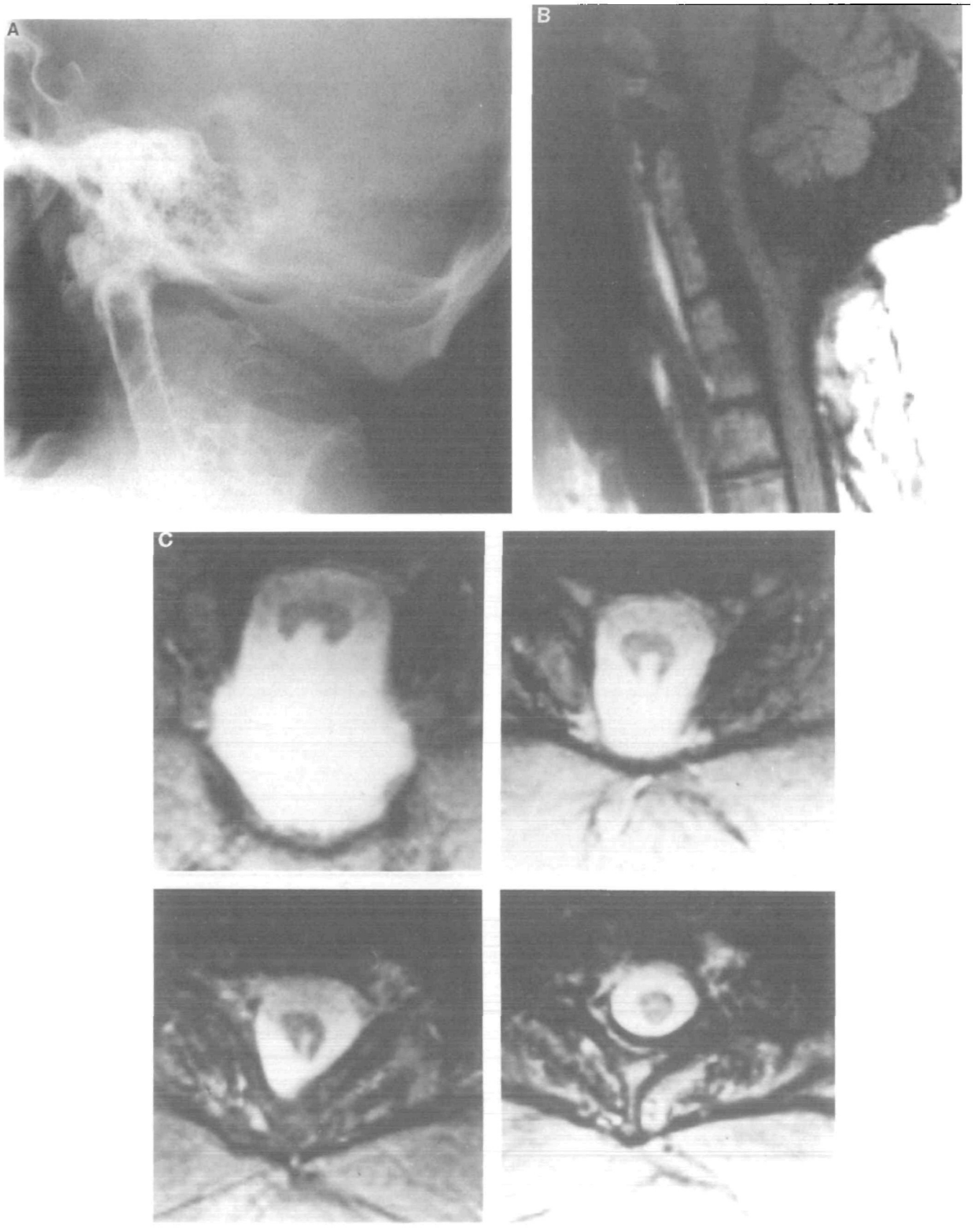

Fig. 6 Case 6. (**A**) Lateral cervical roentgenogram showing extensive fusions between C2 to C6 vertebral bodies. (**B**) Sagittal MRI (T_1-weighted image) and (**C**) axial MRI (T_2-weighted image) with wide foramen magnum, absent posterior vertebral elements and partial posterior splitting of the upper cervical cord.

posterior splitting of the cord was evident at the lower medulla and upper cervical cord (Fig. 6C).

Prolonged pain relief was achieved with injection of local anaesthetic in the area of the left occipital nerve.

Case 7

This 35-year-old lady presented with 10-year history of neck pain. She had had the classic features of Klippel–Feil syndrome since birth.

Examination

Physical examination revealed a short and painful neck with severely limited movements. There were no neurological signs and no visible abnormalities seen over the spine.

Radiological findings

Lateral cervical spine roentgenograms and an MRI scan showed C2/3 fusion and posterior assimilation of atlas to occiput (Fig. 7A and B). The upper cervical spinal canal and the foramen magnum was capacious. An axial MRI scan revealed partial posterior splitting of the upper cervical cord with spina bifida (Fig. 7C).

The patient remained asymptomatic for the last 4 years and her pain was managed conservatively.

Discussion

In 1837 Ollivier reported the first case of sagittal splitting of the spinal cord and named the condition diastematomyelia. It is characterized by sagittal clefting of one portion (rarely more) of the spinal cord, conus medullaris and/or filum terminale (Herren and Edwards, 1940; Lichtenstein, 1940; Bligh, 1961; Naidich, 1983; French, 1990). The spinal cord is divided into two, often unequal sized, portions with either absence or duplication of the central canal and anterior spinal arteries, and one set of anterior and posterior roots per hemicord. The cleft may contain either a sagittally oriented osseocartilaginous spur or a fibrous partition (James and Lassman, 1972; Naidich, 1983). In the former case there are two separate dural tubes while in the latter the split cord lies in a single dural sac.

Diastematomyelia has frequently been associated with vertebral anomalies and dysraphic phenomena (Bligh, 1961; Hilal *et al.*, 1974; Winter *et al.*, 1974; French, 1990). Our cases confirm the observation that cervical cases can often occur with the Klippel–Feil syndrome (Pang, 1992; Ulmer *et al.*, 1993). It has been suggested that patients with cervical diastematomyelia have more extensive vertebral fusions, greater sagittal narrowing of the fused vertebral bodies, as well as greater local widening of the spinal canal than Klippel–Feil patients without split cord (Ulmer *et al.*, 1993). Vertebral fusions and associated bony abnormalities are not, however, inevitable accompaniments of a cervical split cord malformation, as has already been indicated by other reports (Giordano and Davidovits, 1982; Beyerl *et al.*, 1985; Rawanduzy and Murali, 1991).

Suggested developmental basis of the lesions

The seven cases appear to fall into two quite distinct groups. Cases 1 and 2 are characterized by complete splitting of the spinal cord with primarily anterior vertebral defects. We argue later that Case 3 may be a less severe manifestation of this group. Cases 4, 5, 6 and 7 exhibit partial dorsal splitting of the cord with posterior vertebral defects. We argue that these two types of malformation must arise from fundamentally different embryonic lesions.

Complete splitting of the cord with anterior bony defects

Diastematomyelia is most commonly diagnosed in the thoraco-lumbar vertebral region having resulted from disturbance of secondary neurulation in the embryo. In this process the spinal cord forms not by neural folding, as in primary neurulation, but by a process of canalization of a solid spinal cord precursor, termed the medullary cord (Lemire, 1969). Several mutations, and a number of teratogenic insults administered to the vertebrate embryo at the time of secondary neurulation, can yield a split lumbar and/or sacral cord (Cogliatti, 1986; Tibbles and Wiley, 1988; Griffith and Wiley, 1990).

This developmental mechanism, however, cannot explain the phenomenon of split cord in the cervical region, since formation of the neural tube happens here by primary neurulation. Indeed, the cervical/occipital boundary is the site of *de novo* initiation of the entire process of neural folding and neural tube fusion, when the embryo has only 5–7 somites (in the third week post-fertilization of human pregnancy) (Fig. 8A and B). The close proximity of neuroepithelium and notochordal plate during cervical neurulation makes a simple midline lesion, dividing not only the notochord but the neural plate as well, a likely cause of split cord with anterior bony defects. The split notochordal structures then act as organizing centres for split vertebral bodies, separated in the midline by a protrusion of the gut endoderm into the future thecal space (Fig. 8C and D). This provides an opportunity for a neural–enteric connection, as observed in Case 2.

The cervical neural folds appear to have a remarkable intrinsic ability to achieve closure to form a neural tube, as evidenced by the marked resistance to perturbation of this embryonic event in the mouse (Copp *et al.*, 1990). In chick embryos with surgically created midline lesions a single neural tube may form, or the physically separated neural folds may each form a separate neural tube, creating

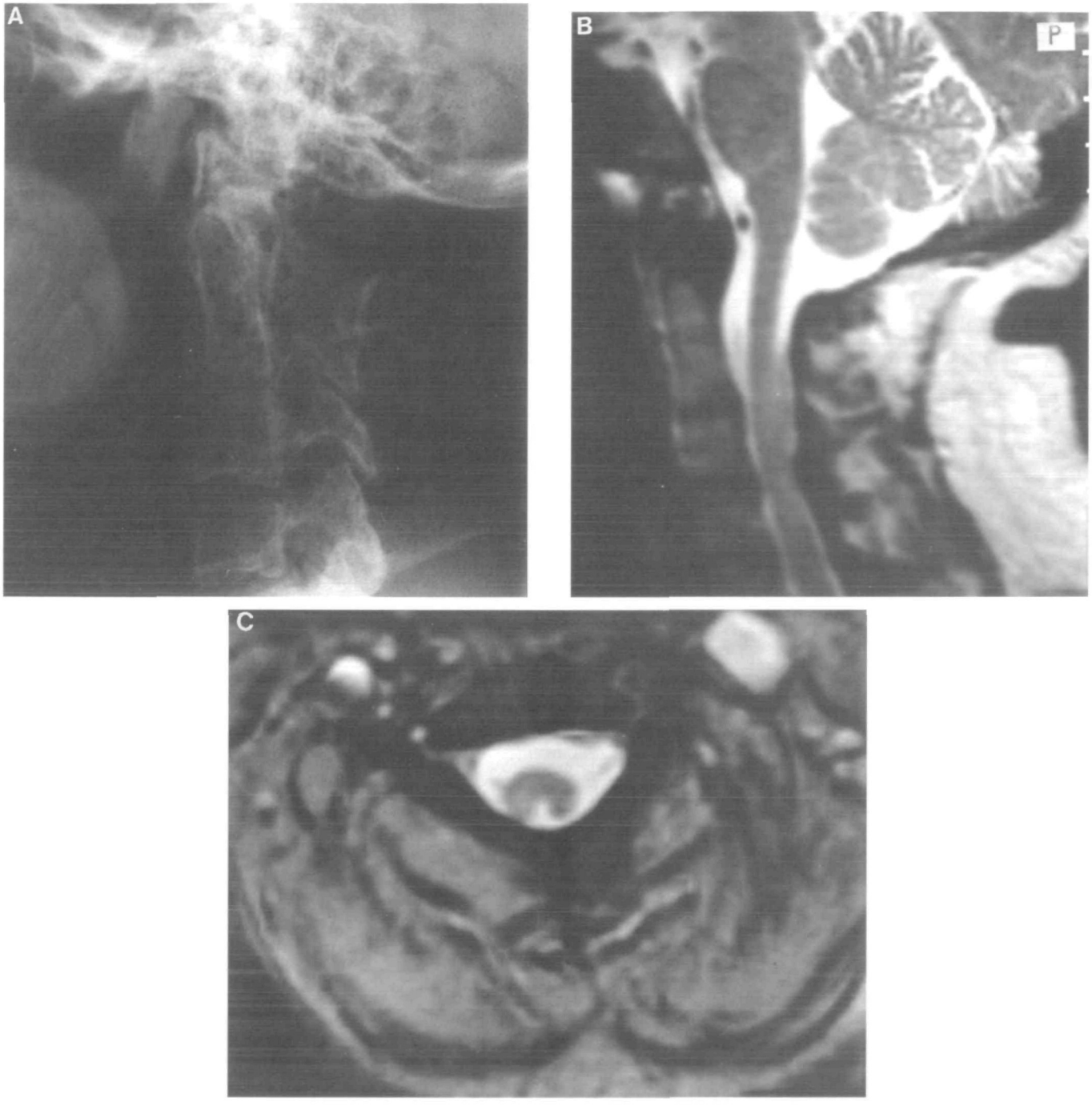

Fig. 7 Case 7. (**A**) Lateral cervical roentgenogram showing partial assimilation of C1 to occiput posteriorly and C2/3 fusion. (**B**) Sagittal MRI (T_2-weighted image) demonstrating capacious upper cervical canal and foramen magnum. (**C**) Axial MRI (T_2-weighted image) with partial posterior splitting of the upper cervical cord.

a split cord (Smith and Schoenwolf, 1991). Others have shown, also in chick embryos, that a simple division of the neural plate does not reproduce diastematomyelia but a firm midline structure is necessary to prevent the fusion of the separated parts (Rilliet *et al.*, 1992).

This suggested pathogenesis is reminiscent of the theory of Bremer (1952) which proposed that the midline abnormality, that bisects not only the notochord but the neural plate at a variable distance from Hensen's node, may be a persistent accessory neurenteric canal. This theory has been recently refined by Pang and colleagues (Pang, 1992; Pang *et al.*, 1992) who suggested that the underlying lesion leading to the formation of an accessory neurenteric canal is adhesion between the endoderm and ectoderm. It is assumed that the abnormal midline fistula forms at about the same time as the (normal) neurenteric canal which is between embryonic days 20 and 24 (Pang *et al.*, 1992; Larsen, 1993). If the mesenchymal condensation

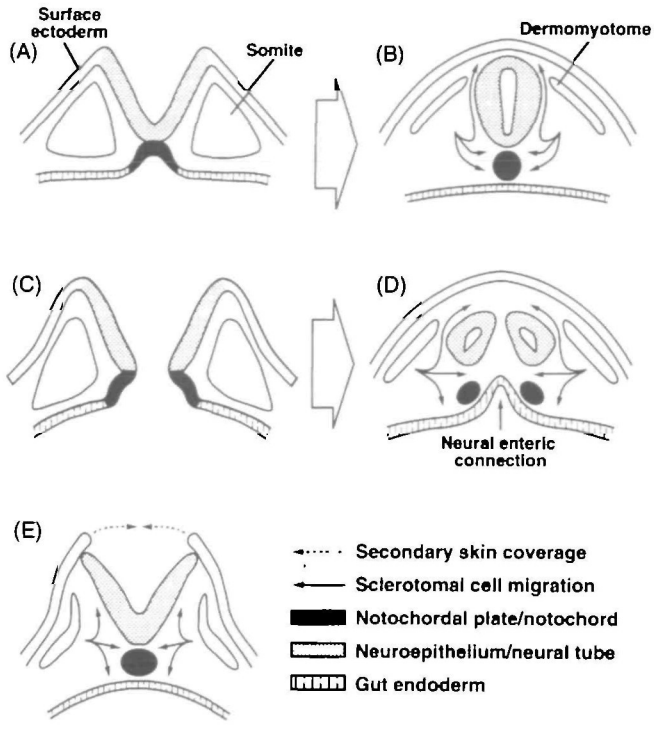

Fig. 8 Suggested embryonic pathogenesis of the cervical defects; (**A**) and (**B**) normal developmental sequence; (**C**) and (**D**) disturbed development as proposed for Cases 1, 2 and 3. (**E**) Disturbed development as proposed for Cases 4–7. Prior to neural tube closure at the cervical level (**A**) the embryo consists of just two cell layers in the midline, the neuroepithelium overlying and directly contacting the notochordal plate. Laterally, the neuroepithelium is attached to surface ectoderm, with their junction comprising the neural fold whereas, ventrally, the notochordal plate is continuous with the prospective gut endoderm. Sandwiched between the dorsal ectoderm and the ventral gut endoderm is the mesoderm which, at this level of the body axis, comprises newly formed somites. As normal development proceeds (**B**) the neural folds elevate and fuse, becoming covered by a continuous layer of surface ectoderm while, simultaneously, the notochord pinches off and comes to lie dorsal to, and separate from, the gut endoderm. Later in development, the ventromedial somitic cells (the sclerotome) lose their epithelial organization and migrate to surround the notochord, forming the vertebral bodies. The sclerotome also surrounds the neural tube, forming the costal and neural processes. (**C**) A midline lesion that splits both notochord and neural plate would be expected to result in a split cord, following independent closure of the separated neural hemi-plates (**D**) together with anterior bony defects as the sclerotomal cells organize around the separate notochordal fragments. A protrusion of the gut endoderm may separate the vertebral bodies. On the other hand, failure of primary neural tube closure (**E**) with secondary soft tissue coverage can yield dorsally split cord, with dorsal bony defects but normal anterior vertebral organization.

finds spatiotemporally available primitive meninx cells at this time in embryonic development they will become incorporated in the midline septum and, due to their sclerogenic potential, may form medial dural walls and an osseocartilaginous septum between the hemicords, resulting in two separate dural tubes. It has been observed that either a fibrous septum or none at all is more common than a bony septum in the cervical cases (Pang, 1992; Ulmer *et al.*, 1993). The normal development of the pachymeninx starts in the ventral side of the thoracic region at around day 30 (Hochstetter, 1934; Sensenig, 1949; O'Rahilly and Müller, 1986). Primitive meninx cells then gradually extend rostrally and caudally and also invest the neural tube. We suggest that these cells are simply not as available in the cervical as in the thoracolumbar region at the time of the midline mesenchymal condensation.

In Case 3, the presence of hemivertebrae and an anterior vertebral defect, in the absence of a posterior bony defect, suggests that a complete septum was present at an early stage of development. However, the persistence of a complete septum can be variable (Pang *et al.*, 1992), offering a possible explanation for the appearance of a partial split cord in this case.

Partial dorsal splitting of the cord with posterior bony defects

Cases 4, 5, 6 and 7 suggest a quite different pathogenesis involving incomplete closure of the cervical neural tube. In the mouse there is only one known genetic cause of failure of cervical neurulation; the *Lp* mutant mouse. In this case, the defect is severe and is propagated into the hindbrain and along the spine, resulting in total craniorachischisis. Human cases of this severe neural tube defect probably result from a similar severe neurulation anomaly (Copp *et al.*, 1994), although the genes responsible have not yet been identified. A less severe, localized disturbance of cervical neural tube closure could be followed by secondary soft tissue coverage perhaps by a mechanism resembling foetal wound healing (McCluskey and Martin, 1995). Sclerotomal cells penetrate dorsally as far as the splayed neural folds, yielding widely spaced rudimentary neural arches, but the open dorsal midline does not become populated by sclerotome, resulting in dorsal midline defects (Fig. 8E).

Vertebral fusions

It is difficult to explain the high incidence of vertebral fusions (either between whole vertebral bodies, hemivertebrae or laminae) observed in these cases. Primary segmentation (correct formation of somites) appears to be controlled by at least two sets of genes. *Hox* genes are needed for correct positional specification within the craniocaudal sequence of somites. Mutations in *Hox* genes, or teratogen-induced disturbance of *Hox* gene expression, causes alterations of the number and identity of cervical vertebrae (Kessel *et al.*, 1990; Charité *et al.*, 1994; Condie and Capecchi, 1994). However, fusions of the vertebrae are not seen in these instances. By contrast, another group of homeobox containing genes, the *Pax* genes may be

involved in primary segmentation: mutations in *Pax-1* yield vertebral fusions affecting the cervical region as well as other levels of the body axis in the mouse (Balling, 1994; Wallin *et al.*, 1994). It seems unlikely that human cases of cervical vertebral fusion involve mutations of *Pax-1* gene; analysis of the paired box sequence of the human *HuP48* gene (most homologous to the mouse *Pax-1* gene) obtained from patients with congenital segmentation anomalies revealed no mutations in this highly conserved sequence (Smith and Tuan, 1994). Nevertheless, the possibility that other genetic and/or environmental influences disturb expression of *Pax-1*, leading to disorders of primary segmentation must be seriously considered.

Evidence that primary segmentation, which normally begins during the third week of gestation, is successfully completed in cases of congenital fusion of cervical vertebrae has been inferred from the normal number of segmental spinal nerves and arteries (Gray *et al.*, 1964). However, during chondrification of the vertebrae, centra elongate at the expense of the intervertebral material and finally fuse, or very nearly fuse, in the region of the perinotochordal area to form an almost solid cartilaginous column for a short time during the eighth week (Peacock, 1951; Gray *et al.*, 1964; O'Rahilly *et al.*, 1983; Müller *et al.*, 1986). Normally these separate laterally at the dense disc anlagen, in turn, converting the cartilaginous rod at this point into a nucleus pulposus (Verbout, 1985). The *Pax-1* gene is strongly expressed in the intervertebral discs of murine embryos during chondrification of the vertebrae. The discs, with their high proliferative capacity, seem to separate the vertebral bodies. In this respect, *Pax-1* gene may function as a resegmentation gene (Wilting *et al.*, 1995). Perhaps this process of forming segmented vertebral bodies and intervertebral discs is erroneous in certain cases due to premature switching off of the *Pax-1* gene, resulting in bony fusion. The relationship between the genes affecting primary segmentation and the division of the spinal cord is not known.

Clinical considerations

The indications for operative treatment of cervical diastematomyelia remain controversial. Spinal cord tethering, that may or may not lead to neurological deterioration, occurs less frequently with cervical diastematomyelia than with the thoracolumbar cases (Eller *et al.*, 1987). One reason for this may be the previously mentioned relative rarity of an osseocartilaginous septum in the cervical cases. The other reason may be that the differential growth rate between the cord and spine (i.e. the relative ascent of the cord) is smallest in the cervical region (Eller *et al.*, 1987; Pang, 1992; Ulmer *et al.*, 1993). Guthkelch (1974) suggested that neurological deterioration could also be due to microtrauma inflicted on the split cord by repeated flexion/extension movements of the neck. However, in the majority of cases, the rigidity of the cervical spine due to its extensive vertebral fusions and capacious spinal canal do not favour this explanation. We believe that these are the major reasons why cervical split cord is usually an asymptomatic and incidental finding, as in our cases in which no radiological and/or clinical evidence of tethering or cord compression was found. Consequently, we agree with others (Okada *et al.*, 1986; Simpson and Rose, 1987; Ohwada *et al.*, 1989; French, 1990; Miller *et al.*, 1993), that prophylactic surgery is not indicated in asymptomatic cervical cases. Close follow-up of these patients is however mandatory and if progressive neurological deterioration occurs surgery must be re-considered.

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