SPINAL DYSRAPHISM*

The Diagnosis and Treatment of Progressive Lesions in Spina Bifida Occulta

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In this paper the results are reported of a study of certain progressive deformities of children's feet which were found to be caused by congenital lesions of the spinal cord and cauda equina, associated with spina bifida occulta. The cases conformed to a pattern and permitted the formulation of a syndrome which has already been described in a preliminary form (James and Lassman 1960). With further experience it is necessary to elaborate the basis of this syndrome and to indicate its validity in diagnosis. We began in 1957 to operate on patients showing this syndrome. In twenty-two out of twenty-four patients operated upon consecutively exploration of the spinal cord has confirmed the diagnosis and has revealed a variety of lesions of congenital origin. Until now no one has had the opportunity to study these lesions. We have classified them according to their apparent mode of action. While we have operated with the intention of preventing further neurological defect and progressive deformity of the lower limbs, there has in fact been some improvement in two out of three patients and no patient has been made worse. The patients have been observed for periods of from six months to four years after operation. We believe that the measure of success that we have achieved is due to relatively early diagnosis. In the past, in patients coming to operation the diagnosis had usually been made too late, when the neurological deficit was extreme, and irreversible vascular and neural changes had occurred.

Our present intention, therefore, is to draw attention once again to the existence of these congenital lesions of the spinal cord and cauda equina, to amplify the syndrome permitting their early diagnosis and to indicate that treatment is possible and worth while. Their incidence is not known, but they are evidently not rare because in seventeen out of our twenty-four cases the diagnosis was made by one of us during the course of routine work. All the patients presented with the syndrome described below, but not all such congenital lesions present themselves in the same manner. We have had experience of other patients in whom the diagnosis was confirmed at operation, who presented with quite a different pattern of physical abnormality. Lesions of the type we are describing have in the past been classified as "myelodysplasia," but this term has been used to cover too many conditions and is no longer specific. We prefer the term spinal dysraphism, which Lichtenstein (1940) revived. This is a better title because it indicates the common origin of the lesions from failure of development in the median dorsal region, and includes abnormalities in the cutaneous, muscular, osseous, vascular and neural tissues, which may occur separately or together. The commonest manifestation is spina bifida. Much has been written on spina bifida aperta but little is known of the lesions associated with spina bifida occulta.

THE CLINICAL SYNDROME

Mode of presentation—The commonest form of presentation is with an established, but slight, cavo-varus deformity of one foot (Fig. 1). The affected leg is slightly shorter than the other. The parents complain that the child walks in an odd manner and twists the forepart of his shoe. There may already be changes in the tendon reflexes and possibly trophic ulceration. In time this cavo-varus foot may develop a paralytic valgus deformity and the other limb may also become involved. Pain is rare. Sensory changes occur, but in children they are almost

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impossible to detect and the findings are unreliable; they should, however, be looked for, particularly in the saddle area of the buttocks.

The whole course of events is the same as with cases of spina bifida cystica, but in spina bifida occulta the process is slower. The speed of deterioration varies from case to case.

The onset of abnormality of the foot may be at any age; some children are born with the early evidence. We have operated on children aged between two and fourteen years. The commonest age of onset seems to be between four and six years. If the first symptom appears in adult life it is usually urinary incontinence and there is no abnormality of the lower limbs.

Diagnosis is easier if there are external cutaneous manifestations on the back—excess hair, a naevus, sacral dimples or a sacral lipoma (Figs. 2 to 5). Eleven of our twenty-four patients had no external manifestations on the back to help with diagnosis; they were diagnosed by observation and examination of the feet and legs.

![Fig. 1](image)

Typical deformity of the foot in spinal dysraphism. Note the cavo-varus deformity.

**Signs**—The earliest evidence in the lower limbs is the gait, which is not unlike that seen when a healthy child has been wearing tight shoes. The abnormal action of the foot in the normal child is transient and soon disappears. This is a point in differential diagnosis at this earliest stage. The lesion of the spinal cord seems to cause an increased neuromuscular irritability of the invertors of the foot so that they overact; the longitudinal arch is elevated and the forefoot adducted. This action is seen only in unselfconscious walking. The muscle imbalance is not otherwise detectable by ordinary clinical examination. The child can control it and there is no detectable loss of power in any of the muscles of the leg and foot. As time passes the muscular imbalance is no longer consciously controllable and the cavo-varus deformity develops.

In some patients deterioration seems to stop when the cavo-varus is established, but they may progress later. In any patient progression can lead to trophic ulceration, incontinence and paraplegia. Such cases have been reported in adults. We have had two children previously paraplegic who are now much improved after operation. One, a boy four and a half years old, has already been reported in detail (James and Lassman 1960). The other patient, a boy aged five, developed paraplegia which was shown at operation to be caused by traction on the conus medullaris by a fibro fatty band passing from the third lumbar neural arch to an intrathecal lipoma situated on the conus. This band was divided and the conus was freed. After some post-operative complications the boy improved steadily and was able to walk again.
Cutaneous manifestations of spinal dysraphism. Figure 2—Excess hair. Figure 3—Sacral dimple. Figure 4—Sacral dimple with pigmented margins and excess hair. Figure 5—Sacral lipoma.
INVESTIGATIONS

Radiography—In every case radiographs will show a spina bifida. A simple split in the spinous process of the first piece of the sacrum by itself can be ignored. Post-mortem dissection and surgical operations have shown us that this bony anomaly is not likely to be associated with a surgically treatable spinal cord lesion.

Myelography—Myelography is essential. The lesion may not be demonstrable by this technique but if it is, its site is indicated.

Our practice now is to introduce the Myodil by the cisternal route with the patient under general anaesthesia. Previously the lumbar route was used but in most of our cases the conus medullaris can be expected to lie at a very low level in the vertebral canal and the spinal cord may be damaged by insertion of the needle. No injury of this kind has yet occurred, but we recognise the risk. Also, some of the Myodil may be injected or leak into the subdural (or extrathecal) space and obscure detail in subsequent screening. This has occurred in several of our cases so that the patient has had to be submitted to the examination for a second time some months later. Even so, the misplaced contrast medium has not been completely absorbed. Myodil escaping outside the arachnoid after cisternal puncture is unlikely to interfere with this examination. Whitby (1961) has reported the anaesthetic technique used for our cases at myelography.

Dr G. Gryspeerdt has been responsible for the myelographic examinations of our patients and has developed an important technique for demonstrating the level of the conus medullaris (Gryspeerdt 1961). In the absence of any demonstrable lesion, when the conus lies at the level of the third lumbar vertebra or lower, there will be a lesion in the region of the conus and cauda equina.

In a previous paper (James and Lassman 1958) we noted that there was no published information about the level of the conus medullaris between birth and adult life. The results of our examinations of necropsy specimens suggest that the spinal cord has completed its ascent within the vertebral canal by the age of five years and that at this age the conus medullaris normally lies at the level of the upper border of the body of the second lumbar vertebra.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis is wide. The most important conditions to exclude are spinal neoplasms, Friedreich's ataxia, cerebral palsy, old poliomyelitis and local conditions of the feet. We have erred in our diagnosis in two of our twenty-four patients. One proved to be a case of Roussy-Levy syndrome and the other is still undiagnosed.

THE LESIONS

Diastematomyelia—In the last few years a considerable amount of attention has been paid to diastematomyelia—a bifid state of the spinal cord of developmental origin. This is only one of several congenital anomalies of the spinal cord and its coverings which produce similar clinical findings. We wish to emphasise that where diastematomyelia is suspected and not found by radiological methods, other congenital anomalies affecting the spinal cord must be sought.

In diastematomyelia there may or may not be a septum passing dorso-ventrally between the two halves of the spinal cord. This septum may be fibrous, cartilaginous or osseous. The bifid cord is not by itself necessarily the cause of symptoms; they are produced by the septum or other associated lesion.

The lesions affecting the spinal cord are extrinsic and for general purposes can be classified in three groups according to their supposed mode of action.
Lesions causing traction—These are the commonest and they act by preventing the so-called ascent of the spinal cord during the course of growth in length of the vertebral column. The spinal cord becomes tethered to the subcutaneous tissues, to the vertebral column or to the dura mater. The tethering agent may be an ectopic posterior nerve root, arachnoid adhesions, fatty fibrous adhesions, an abnormally developed filum terminale presenting as a fibrous band attached in an abnormal position, or a fibrous band passing through to the subcutaneous tissues. A septum associated with diastematomyelia may also produce a traction lesion.

<table>
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<th>Operation finding</th>
<th>Number of patients</th>
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<td>Lesion causing pressure</td>
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</tr>
<tr>
<td>(includes one case of diastematomyelia)</td>
<td></td>
</tr>
<tr>
<td>Lesion causing traction</td>
<td>9</td>
</tr>
<tr>
<td>Lesion causing pressure and traction</td>
<td>11</td>
</tr>
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<td>(includes ten cases of diastematomyelia)</td>
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<tr>
<td>No abnormality</td>
<td>2</td>
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Lesions causing pressure—These may be: transverse bands of ligamentous origin, usually extradural; inverted lamine; intrathecal lipomata; fatty fibrous tissue associated with defective neural arches, either intrathecal or extradural or both; intrathecal dermoid cysts, or neurofibromata. Septa associated with diastematomyelia may also cause pressure.

Lesions causing traction and pressure—These may be any of the above in combination. They may act together at the same time or either the pressure or traction effect may be exerted separately at different times. It is often impossible to decide which is the cause of the damage.

TREATMENT

Treatment consists of laminectomy, exposure and opening of the dura mater, and exploration of the spinal cord. Lesions preventing or likely to prevent ascent of the spinal cord or causing pressure are removed. The old notion of a tight filum terminale has a basis of truth, and exploration in the lumbo-sacral region should include examination of this structure. The dura mater must be opened in every case: many extradural lesions are found and many of them continue within the theca; these must be removed and the spinal cord be seen to have no tethering agent.

We undertook operation with the chief object of preventing deterioration. However, in two-thirds of the patients some degree of improvement followed operation.

CASE REPORTS

Six illustrative cases from our series of twenty-four are described below; the case of a seventh patient, not in the series because the operation was done only recently, is added to illustrate rapid healing of a severe trophic ulcer after operation.

Case 1—A girl aged five years and nine months was brought because of “cavo-varus” gait with three-quarter inch shortening of left leg and one inch shortening of left foot. The latter had been first noticed by the mother shortly after birth. There was diurnal incontinence of urine.

The shape of the left foot was normal but there was a tendency for the heel and forefoot to invert on standing and more so on walking. The left ankle jerk was absent. There were no detectable sensory changes. A sacro-coccygeal dimple was present.
Seven months later she had developed some adduction of the left forefoot which was not passively correctable; her condition otherwise was unchanged.

Radiographs showed spina bifida from the fourth lumbar vertebra down to the lower sacral region. The distance between the pedicles was greatest at the first sacral level (Fig. 6). Myelography showed that the theca was held posteriorly in the lower lumbar and sacral region. The conus was at the fourth lumbar level and the subarachnoid space ended at the second sacral.

At operation—laminectomy from the fourth lumbar to the fourth sacral vertebra—an extradural traction band was found to pass inwards—probably from the dimple—entering the theca at the second sacral level and passing along the left side of the filum terminale to attach to the conus at the level of the caudal border of the fourth lumbar vertebra (Figs. 7 and 8). The dural sac continued to the fourth sacral level. The band and the filum terminale were divided, to leave a gap of one centimetre in the latter. Histological examination showed the band to be a nerve.

Two months after operation the child's gait was normal but the deformity of the foot was unchanged. The ankle reflex was still absent and control of the bladder was still defective.

Nineteen months later the only further change was a possible improvement in bladder control.

Case 2—A girl aged six years and four months was brought because she walked on her left forefoot, her left knee did not appear to straighten completely when she stood, and because there was half an inch of shortening of the left leg and foot. She had not used her left leg properly when she first started walking and at the age of eighteen months she had seen a consultant who recognised the existence of the spina bifida occulta and noted the presence of a sacro-coccygeal dimple. He had advised wedging of the shoe to correct the tendency to inversion of the foot. Walking apparently improved but the child was lost sight of.

The only overt abnormalities were equinus position of the foot on walking and the shortening of the left lower limb, which was mostly in the leg. Later, the foot tended to assume a valgus position and developed slight cavus deformity. When the girl was seven years eight
months old clawing of the toes was noticed. Tendon reflexes at the knee and ankle were always difficult to elicit and variable in briskness, but the right were easier to obtain than the left. The left plantar response was extensor for some years; the right was flexor throughout. No sensory changes were detected at any time. The left leg was occasionally more cyanosed and colder than the right.

Radiographs showed spina bifida of the tenth to twelfth thoracic and of the fifth lumbar and first sacral vertebrae. The body of the eleventh thoracic vertebra was represented simply by a forward extension from each pedicle (Fig. 9).

Myelography suggested diastematomyelia at the eleventh thoracic level, where there was also a temporary hold-up of the flow of Myodil. The conus was abnormally low.

At operation—laminectomy of the tenth to twelfth thoracic vertebrae—an extradural traction band was found passing caudally, possibly to the dimple; the band passed internally through the dura at the level of the eleventh thoracic neural arch and changed direction caudally to attach to the spinal cord at the caudal end of the diastematomyelia at the level of the twelfth thoracic neural arch. There was diastematomyelia three centimetres long without a septum. The conus was not seen. The traction band was divided both intradurally and extradurally, and the sacro-coccygeal dimple was excised (Figs. 10 and 11).

Histological examination showed that the traction band was a fairly large nerve, the intradural portion being connected to a posterior root ganglion.

Five weeks after operation there was subjective improvement in the mobility and warmth of the left foot and toes. The gait was as normal as would be expected with the degree of shortening present.

Ten months after operation the gait was normal and the foot deformity minimal. Shortening remained the same. Tendon reflexes were unchanged.

Fourteen months later there was no further important change, though there was a hammer-toe deformity of the middle digit. In this case there was some subjective improvement, some slackening of the clawing of the toes and some lessening of valgus deformity.

Case 3—A boy was first seen at the age of five months because the left leg was thinner and the left foot smaller than the right. There was no other clinical abnormality. When the boy was nine months old his mother thought he might have some diminution in the sensibility of the left lower limb. At that time too hair was beginning to grow in the lumbo-sacral region. Spina bifida occulta was then diagnosed. The boy started walking on his own at fourteen months, but both feet showed such valgus deformity that he was given arch supports. When he was four and a half years old it was thought that his left tibialis anterior muscle might be paralysed and slight shortening (about quarter of an inch) of the left lower limb was found. When the boy was eight his left tibialis anterior muscle was working strongly and his mother noticed the beginning of inversion of the heel and adduction of the forefoot. This slowly became more obvious clinically. Both mother and boy thought sensation in the left foot was defective but this was not objectively demonstrable. If he developed a sore place on the foot it was slow to heal.
At eight years four months the left lower limb was half an inch shorter than the right and the foot at least one and a half shoe sizes smaller than the right. The tendon reflexes in the right lower limb were all normal; the left knee jerk was absent and the left ankle jerk was very difficult to obtain. It was impossible to test the left plantar response owing to the marked clawing of the toes. The left heel was inverted and the forefoot adducted. Myelography was performed but the appearances were regarded at the time as insufficiently abnormal to justify laminectomy.

During the next ten months he wore an instrument to control the strong inversion action of the foot, but in spite of this the shoe became badly distorted. The tendon reflexes in the left lower limb were definitely absent by the time the decision to operate was made. The right lower limb remained normal throughout.

Radiographs showed spina bifida of the whole lumbar spine and sacrum. Myelography showed that the theca was widened in the middle and lowest lumbar region and had an increased antero-posterior diameter. The appearance of the filum terminale was abnormal and in consequence the conus was not identifiable.

At operation—laminectomy from the second to the fifth lumbar vertebra—the filum terminale was found to be attached over a length of one centimetre by strong fibrous adhesions to the dura mater on the right side at the level of the fifth lumbar vertebral body. The filum continued caudally to the second sacral level and from the conus to the adhesions was suspended from the posterior dura by a vertical veil of arachnoid (Fig. 12). The conus was at the third lumbar level. The adhesions and arachnoid veil were divided and the filum was cut across to leave a gap of 2.5 centimetres.

Histological examination confirmed the identification of the filum terminale. The arachnoid veil was identified as fibrous tissue.

Four weeks after operation the boy was wearing ordinary shoes normally. The gait, allowing for the deformity, was approaching normal. The left buttock was felt by the boy to be "numb," but was hypersensitive. Twenty-one months later no sensory abnormalities were detectable. The deformity of the foot was possibly a little more evident; there was callosity over the base of the fifth metatarsal and there was clawing of toes. The left ankle jerk was present but diminished; the other reflexes were unchanged. Shortening remained less than half an inch.

Case 4 (Mr J. B. Kyle's case)—A boy was first seen at the age of two years nine months because his left leg seemed thicker than the right. There was a difference of half an inch in the circumference of the lower thighs. The only other clinical abnormality was hypertrichosis in the lower lumbar region. He was lost sight of for four years and was seen again because of persistent equinus gait with the right leg. The tendon calcaneus was tight and the right calf muscles felt spastic. The right plantar response was extensor. At eight years nine months, the right foot was seen to be slightly shorter than the left. At nine years eleven months the right leg was half an inch shorter than the left and at ten years four months the disparity was about one inch. At this time there was "clasp-knife rigidity" in the right lower limb; the knee jerks were brisk and equal; the left ankle jerk was brisk but the right was only obtained with difficulty; the left plantar response was flexor and the right extensor; there was no detectable sensory loss. The feet appeared normal except for a tendency for the toes of the right foot to hyperextend.

Radiographs showed spina bifida from the third lumbar vertebra to the second sacral segment (Fig. 13). The appearance on myelography
suggested that in the lumbo-sacral region the theca was rather more posteriorly placed than usual. The conus was at the fourth lumbar level.

At operation—laminectomy from the third lumbar to the first sacral vertebra—a traction band was found extending from the fatty fibrous tissue in the area of the spina bifida to the dura, entering the theca and then fanning out into a series of bands which attached to the conus at the level of the cranial border of the fourth lumbar vertebra and held the conus and cauda equina posteriorly (Fig. 14). The bands were divided and the conus and nerves fell to lie normally against the vertebral bodies.

Histological examination of the traction band showed it to consist of dense fibrous tissue in which was embedded a small nerve.

Five weeks after operation the gait was normal and there was no evidence of spasticity. The reflexes were unchanged and the toes still tended to hyperextend.

Eighteen months later the condition remained unchanged.

Case 5—A girl was first seen at the age of ten years because her right leg was one inch shorter than the left; the right foot was shorter than the left and there was early pes cavus. There was hypertrichosis in the upper lumbar region. The plantar responses were flexor but the right knee jerk was diminished and both ankle jerks were absent. Six months later the right knee and ankle jerks were absent and the left were normal; the left plantar response was flexor, but there was no definite response in the right foot. Vibration sense over the right medial malleolus was impaired and light touch sensation was absent on the outer aspect of the right foot. Pinprick however could be felt.

Radiographs showed that there were eleven thoracic and six lumbar vertebrae. There was spina bifida in the lower thoracic and upper lumbar regions and to a lesser extent in the lower lumbar region. The bodies of the tenth and eleventh thoracic vertebrae were fused. Tomography showed a bony septum extending from the neural arch of the ninth thoracic vertebra to the posterior aspects of the bodies of the eighth, ninth and tenth thoracic vertebrae.

Myelography showed an apparent space-occupying lesion—in fact, a diastematomyelia—in the lower thoracic region. The level of conus was not determined, but it was probably at the third lumbar level. The flow of Myodil was constantly held up on the right side at the caudal part of the bony septum, and the contrast medium tended to pocket there.

Operation was undertaken when the girl was ten years and seven months old. The laminae of the eighth to the tenth thoracic vertebrae were removed. A bony septum was continuous antero-posteriorly with the neural arch of the ninth thoracic vertebra, whose laminae compressed the dura on either side. The spinous process of the ninth thoracic vertebra had a hole in it through which passed a round, probably fibrous, band which was attached to the dura. No intrathecal continuation was found; the band probably merged with the bony septum. The spinal cord was firmly adherent to the sides of the bony septum and had to be separated by sharp dissection of the arachnoid adhesions. There was localised arachnoiditis over the spinal cord at the level of the bony septum. There was diastematomyelia over a length of 6-8 centimetres. The bony septum was excised (Fig. 15).

Histological examination of the bone showed it to contain marrow cavities. The adhesions were confirmed as being thickened arachnoid without evidence of inflammatory cell infiltration.
Ten months after operation the right knee reflex was present but diminished. There was no increase in the deformity of the foot.

Eighteen months later the knee reflexes were equal. If allowance was made for the disparity in length of the lower limbs the gait approached normal.

**Case 6**—A girl born by breech delivery was first seen at the age of one month because she had a left foot drop which was apparently recovering. The left calf was slightly smaller than the right and there was a suspicion of left pes cavus. The weakness of the foot recovered completely but when the child was a year old the left foot was smaller than the right and had a strong tendency to inversion which became worse over the next three months to produce an apparent equinovarus deformity. By the time the child was twenty-one months old the equinovarus tendency had ceased and progressive valgus deformity occurred, to become established over the next fifteen months. At that time the forefoot was pronated, the outer toes were abducted and the hallux was held in valgus. There was defective circulation in the toes which were frequently septic, particularly the little toe. When the girl was four the right plantar response was flexor but the left was unobtainable and there was absence of superficial sensation in the left lower limb. The mother commented at this time that smacking the child on the left buttock had no effect, so she always smacked the right. About this time the left ankle jerk was noted to be absent. There was no disparity in length of the lower limbs.

Radiographs showed a bony septum at the twelfth thoracic level with spina bifida of the eleventh thoracic, of the first, second and fifth lumbar vertebrae and of the sacrum. There was also an anomalous development of spinous processes and laminae with spina bifida of the first and second thoracic vertebrae (Fig. 16).

![Figure 16](image1.png)

**Fig. 16**

Case 6. Figure 16—Radiograph showing spina bifida of the eleventh thoracic and first, second and fifth lumbar vertebrae and of the sacrum. There is a median bony septum at the twelfth thoracic level.

![Figure 17](image2.png)

**Fig. 17**

Figure 17—Myelograph (prone position) showing the defect caused by the bony septum.

![Figure 18](image3.png)

**Fig. 18**

Case 6—Findings at operation. Figure 18—Extradural appearances. The bony septum is seen centrally, with the dura mater cranial and caudal to it. The latter is bulging and shiny in comparison with the healthy dura cranial to the septum. Figure 19—The dura mater has been opened and the bony septum and surrounding dura mater have been removed. The septum was closely adherent to the caudal junction of the two cords, and the caudal part of the cord appears engorged with blood in comparison with the cranial portion.

Myelography confirmed diastematomyelia at the twelfth thoracic level. The level of the conus was not determined (Fig. 17).

Operation was performed when the girl was six years three months old. The laminae of the twelfth thoracic and first lumbar vertebrae were removed. That of the eleventh thoracic vertebra was
absent. The bony septum was localised and the dura caudal to it was seen to bulge considerably in comparison with that on the cranial side. The septum was removed. When the dura had been opened the bifid spinal cord caudal to the level of the septum was seen to be swollen and engorged with blood. The junction of the bifid spinal cord had been very tightly applied to the bony septum. The length of the diastematomyelia was 2.5 centimetres. The conus was at the level of the second lumbar vertebra (Figs. 18 and 19).

Six weeks after operation the child felt her legs to be stronger and was able to hop on either foot. Subjectively there was improvement in sensibility. There was less evidence of cutaneous circulatory disorder.

At twelve months the left ankle jerk was still absent and there was no change in the deformity of the foot. The girl was more active and felt the left leg to be stronger. Septic lesions of toes were occurring less frequently.

![Case 7—Trophic lesion of great toe before (left) and six weeks after operation.](image1)

**Case 7 (Professor S. D. Court’s case)—A girl aged six and a half was seen because of persistent sores on the toes of the left foot which had started about nine months previously. The left foot had always been thought to be slightly shorter than the right. There was early cavovarus deformity of the left foot and there was a lipoma over the sacrum with a shallow dimple about one inch in diameter at its lower margin. The left knee and ankle reflexes, the right ankle reflex and the left plantar response were all absent. There was a severe trophic lesion of the great toe (Fig. 20).**

Radiographs showed spina bifida from the second lumbar vertebra downwards including the whole sacrum (Fig. 21). At myelography the contrast medium flowed to the left side of the vertebral canal in the lower lumbar region and then entered a cavity which was a caudal continuation from the terminal thecal sac, extending cranially and anterior to the subarachnoid space around the spinal cord. This sac displaced the main column posteriorly, but all the contrast medium became trapped in it so that further examination became impossible.

At operation—laminectomy of the fifth lumbar and first and second sacral vertebrae—an extradural traction band was found at the caudal border of the second sacral lamina. It was connected to the subcutaneous tissues and was attached to the dura at the level of the first sacral lamina. The band passed intrathecally and joined the conus at the level of the caudal border of the first sacral body. The band was excised. The filum terminale was wide and was attached more caudally than normal (possibly to the fourth sacral vertebra). On its left side at the level of the second sacral lamina there was a very small opening into the sac which had been seen on myelography and which was seen to be a sacral arachnoid cyst. The filum did not appear tight but when it was divided the ends retracted to leave a gap of 2.5 centimetres (Figs. 22 and 23).

Histological examination showed that the extradural band was fibrous tissue with cartilage and bone.
After operation there was rapid improvement in the trophic ulceration of the left foot, but no other change (Fig. 20).

DISCUSSION

It is too soon to be certain that operation in cases of occult spinal dysraphism will prevent further neurological defect, but our results suggest that laminectomy is justified. The operation is often extremely difficult technically and should be done only by surgeons who are already familiar with the surgery of the spinal cord. Although the lesions themselves fall into a set of broad patterns and may be dealt with as such, the variations in each case require careful consideration in detail.

Fig. 22

Case 7—Findings at operation. Figure 22—The dura mater has been opened. The subcutaneous tissue and the fibrous band leading from it to the dura are being retracted. The extradural continuation of the band passes behind the two central stay sutures. The retracted dura mater reflects the light strongly and appears convex, though it is actually concave. The band is seen again to the left of the retracted dura, fanning out intrathecally to be attached to the conus medullaris at the first sacral level. The filum terminale is running in the midline in the centre of the field, but passes towards the right in the right part of the field. Figure 23—Appearances before closure. The fibrous band has been divided at its points of attachment, which are seen projecting just to the left of the centre of the field. To the right of centre the cut end of the filum terminale and the gap produced by its retraction are clearly shown. The original point of attachment is the blackened area seen at the caudal (right hand) end of the dural opening.

If operation is to be successful, diagnosis must be made early before irreversible changes have occurred in the spinal cord or cauda equina. Neurological surgeons will not see these patients early unless the orthopaedic surgeons and paediatricians make the diagnosis.

At first we used myelography only in cases where we considered that laminectomy was indicated but we now use myelography for diagnostic purposes in doubtful cases. In Dr Gryspeerdt’s experience of the use of Myodil in 1,400 examinations over ten years there has been no serious complication.

In general, our criteria for laminectomy are: 1) Abnormality of the gait and deformity of the foot either progressive or associated with neurological defect. 2) The presence of spina bifida of grosser degree than a simple split of the spinous process of the first sacral vertebra. 3) Myelographic evidence of abnormality or of a low conus medullaris in patients suspected of having a lumbo-sacral or sacral lesion.

Our experience covers a wider field than the twenty-four cases on which this paper is based and we have come to the conclusion that preventive operation is indicated in some patients who simply have external cutaneous manifestations on the back. The incidental finding of diastematomyelia with a bony septum may not by itself be an indication for operation; in our experience the condition only came to light because of abnormality of the lower limbs. During our studies on the level of the conus in the cadaver we have seen an instance of diastematomyelia in a person of sixty-seven. The double cord was transfixed by a bony septum at the fifth lumbar level, but there had been no neurological abnormality or deformity.

Deformity of neurological origin of one or both feet can cease to progress after operation. It may be possible, in young children, for a foot deformity to progress during growth because
the bones of the feet are already deformed and not because the neurological lesion is still operative. This does not however seem to apply to older children. Some of our patients had such marked deformity before operation that local treatment was evidently going to be necessary later. Others, with a small degree of deformity which had been expected to need local treatment, have improved after operation and are now unlikely to require further local treatment.

Since all these patients have a neurological defect of some kind, it should in theory be possible to make an accurate assessment on clinical examination. In practice we have found that with tests of sensibility requiring subjective response, most children gave answers so contradictory and variable as to make the tests valueless. Positive information was obtained only in patients who had complete anaesthesia or trophic ulceration, although we considered that those children with superficial evidence of poor circulation in the lower limbs might also have sensory loss. Electromyography was done in some cases but the results were not helpful and the tests were stopped because they were unpleasant for the children.

SUMMARY

1. A syndrome resulting from congenital lesions affecting the spinal cord and cauda equina, associated with spina bifida occulta, is described.
2. The syndrome consists of a progressive deformity of the lower limbs in children. One foot and the same leg grow less rapidly than the other. The foot develops a progressive deformity which begins as a cavovarus and becomes a valgus one. Both lower limbs may be affected. There may be progression to sensory loss, trophic ulceration, disturbance of function of bowel and bladder and even paraplegia.
3. Methods of investigation including myelography are described.
4. Exploration of the spinal cord has been undertaken in twenty-four patients so affected. Extrinsic congenital lesions causing traction or pressure or a combination of traction and pressure on the spinal cord have been found in twenty-two of these.
5. In two-thirds of the patients some degree of improvement has followed operation.

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REFERENCES


