SPINAL DYSRAPHISM

AN ORTHOPAEDIC SYNDROME IN CHILDREN ACCOMPANYING OCCULT FORMS

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Much interest has been shown in the pathology of the more gross developmental anomalies of the spinal cord and its coverings and in their clinical manifestations, most of which are not amenable to treatment. It has not been appreciated that lesser anomalies can also produce disabilities, that they can be diagnosed in life and that they can frequently be treated by surgery before the secondary effects have become severe and irreversible. Clinical experience over a number of years of the many children sent to orthopaedic clinics with various types of foot or lower limb defect led us to suspect the presence in some children of a spinal cord lesion responsible for a particular pattern of lower limb abnormalities. Such children were watched carefully and the development of their abnormalities and subsequent radiological and neurological investigations confirmed us in our belief that in a number of those children presenting at first with relatively trivial foot defects, there was an underlying spinal anomaly causing their troubles which might well be dealt with at an early stage, thus preventing further disability and perhaps improving such disability as had already occurred. We therefore decided to operate, at first upon those children whose abnormalities were beginning to increase seriously, and later, in the light of experience gained, upon all those in whom we had good reason to suspect the presence of a spinal lesion.

We now believe that we can identify with certainty a syndrome associated with and diagnostic of spinal dysraphism when there is an occult lesion affecting the spinal cord. By diagnosing this condition early we can undertake spinal surgery before severe deformity and disability of the lower limbs has occurred confident that surgery will be beneficial. The clinical manifestations are for the most part orthopaedic in that it is the lower limbs and feet

which usually suffer. The treatment in the first place is principally in the field of neurosurgery since the removal of the primary cause, when it is possible, demands laminectomy and exploration around the spinal cord within the dura mater. Subsequent orthopaedic care will be needed only to correct established deformity if the diagnosis has been made late.

Spinal dysraphism is a term which has been applied to failure of complete development in the midline of the dorsal aspect of the embryo. The extent of this failure may be of mild, moderate or severe degree. The best known clinical form is meningocele which is an example of an overt lesion, but the lesion may not be visible on the surface nor make itself evident clinically. Spina bifida occulta is such a condition and is so commonly noted in routine x-ray investigations that it is regarded as a curiosity rather than as an entity with possible clinical implications. Ingraham and Lowrey (1943) have given an indication of the types of lesion to be found clinically, and other writers have described the particular lesions which interested them. None appears to have regarded these lesions as being of the same origin except Lichtenstein (1940), who revived the term spinal dysraphism and under this heading grouped disorders according to whether they manifested themselves as arising from cutaneous, mesodermal or neural derivatives of the dorsal median region of the developing embryo.

In this communication we are concerned only with those forms of spinal dysraphism which directly or indirectly affect the spinal cord and spinal nerves and our aims are to describe these lesions, to indicate the general pattern of the associated orthopaedic syndrome and to draw attention to the value of spinal surgery as a preventive measure when undertaken early.

The Lesions

Lesions affecting the spinal cord or cauda equina may be classified into three groups, those which produce a traction effect, those which exert local pressure and those which combine traction and pressure.

The Traction Group. In the growing child, there is a difference between the rate of growth of the spinal cord and that of the vertebral column. This is described as the ascent of the spinal cord within the vertebral canal, because the length of the latter, as the vertebral column grows, increases at a greater rate than the length of the spinal cord. At birth the distal end of the spinal cord is normally situated at or about the upper border of the body of LV.3 and at the end of growth, in adult life, is situated at the lower border of the body of LV.1 or the upper border of LV.2 (Streeter, 1919). Any lesion which prevents this ascent of the spinal cord will do so by fixing it at a definite level in the vertebral canal. In such circumstances the spinal cord or nerve roots or both will be stretched as the bone column grows and this stretching will produce injury either to the tracts within the spinal cord, or to the nerve fibres and indirectly to both of these by interfering with their blood supply. In all probability the commonest lesion of this type is diastematomyelia in which the spinal cord is bifid in part of its course. This is also the most easily diagnosed lesion when it is associated with a bone septum lying anteroposteriorly across the vertebral canal and transfixing the spinal cord. This bone septum can be seen on a plain x-ray film. A septum is, however, not always calcified; it may be fibrous and therefore not demonstrable by plain x rays.

Fibrous bands are also found tethering the spinal cord or cauda equina to the bone or skin directly or through attachment to the meninges. These may be associated with the fibrolipomata found in the pressure type of lesion. A tight filum terminale has often been cited as a similar traction lesion.

The Pressure Group. The best known type is the intrathecal lipoma, which consists of a fatty fibrous mass lying inside the dura mater. In some cases the fatty fibrous mass is entirely extradural; in others it may be extradural and intrathecal. Another type of pressure lesion occurs when a fibrous band constricts the lower end of the spinal cord situated at an abnormally low level (Léri, 1926). Occasionally, when the vertebral canal is already narrowed by inversion of one or more laminae, pressure may arise from increasing thickness of the bone with normal growth.

Within this group may also be placed those cases

of congenital scoliosis resulting from hemivertebrae which develop paraplegia at about the age of 12 years.

The Combined Group. This group consists of a variety of lesions which incorporate external pressure by a fatty fibrous mass or by a transverse band with traction by some form of fibrous band attaching to the meninges, bone or skin.

Symptoms and Signs—Early Stages

Occult forms of spinal dysraphism are rarely evident at birth. At that time, the function of the spinal cord may not be impaired, but as the child grows, a dysraphic lesion will begin to have an effect, either by preventing the spinal cord from 'ascending' within the vertebral canal or by pressure from the increasing size of some abnormal tissue such as a lipoma. The effect, then, of a dysraphic lesion is a gradual interference with conduction in the spinal cord or its nerve roots, so that according to the site of the lesion there will be a gradual loss of sensation and a slowly developing weakness of some muscles leading to muscle imbalance and ultimately to deformity.

Symptoms. A child of any age is commonly referred because of a peculiarity of gait or because of distortion of one shoe, itself the result of a peculiarity of gait unnoticed by the parents. Sometimes a child attends because of poor posture associated with a short leg or because one leg has been noticed to be shorter and thinner than the other.

Pain is not a feature but there is occasionally a complaint of pain in one or more metatarsophalangeal joints which may be associated with a protective kind of gait. This is possibly the earliest evidence of this syndrome since in such cases at this stage there is no foot deformity.

Gait. The abnormality of gait, almost imperceptible early on, consists in an elevation of the first metatarsal head as though something like a drawing pin were underneath it, the great toe usually remaining flexed. Occasionally this is accompanied by adduction movement of the fore-foot, but there is no evidence of abnormal function in the foot or toes when standing or when not bearing weight; voluntary actions are normal. In the course of review the peculiar action is more easily seen and the deformity which follows is quite obviously the result of this peculiar action; even so, the foot may still have a normal appearance at rest. If allowed to continue, the pes cavo-varus observed only when walking becomes fixed. The arch

FIG. 1a

FIG. 1b

FIG. 1c

becomes higher, the forefoot more adducted and more inverted, and the toes become clawed. The result (Fig. 1 a, b and c) is not unlike the deformity seen in some cases of poliomyelitis and cases of relapsed club foot.

Lower Limbs. Examination of the lower limbs almost always reveals a shortening of one leg and foot, although it may be absent or very slight in the early stages. The affected leg is smaller in girth, both in the thigh and in the calf. The shortening of the leg rarely progresses to a final discrepancy of as much as 2 in. and it is usually much less, but the shortening of the foot becomes exaggerated by the cavo-varus and clawing of the toes. Peripheral circulatory deficiency is sometimes evident, showing as a lividity or cyanosis.

In fully developed cases the combination of muscular imbalance and ischaemia produces fixed clawing with atrophy of the toes.

Nervous System. Reflexes are usually normal but may progress to an extensor plantar response and exaggerated, and later, absent knee and ankle jerks, as may occur in all progressive neurological lesions. Sensory changes have not been detected in younger children owing to their unreliable testimony, although some have exhibited trophic

FIG. 1.—Case 6. The deformity of the left foot in a moderately severe case. Right foot normal.

- a Showing the inversion of the forefoot and the degree of cavus. Early clawing of the toes. Standing, bearing weight.
- b Showing the inversion of the heel. The thinning of the calf is not well shown. Standing, bearing weight.
- c Showing the shape of the sole and the shortening of the foot. Kneeling; the feet are not touching the background.

changes. Older children manifest the usual alterations in sensation in the leg through dulling of light touch and hypersensitivity on deep pressure to total loss of sensation and also the characteristic saddle shaped area of analgesia over the buttocks and posterior surface of the thighs. We have not tested for loss of anal tone which other authors have found to occur (Jones and Love, 1956). Testing sensation in any child is difficult because the child will often give the answer which he thinks the examiner wants. The results are therefore commonly unreliable.

Back. There is frequently no superficial evidence of abnormality in the spinal column; occasionally a bifid spinous process may be palpable. The visible abnormalities in the back are cutaneous or subcutaneous; they are not present in every case and when present are not always associated with neurological deficit or foot deformity although these may develop later. They consist of the following:

(1) *Subcutaneous lipoma.* This is often associated with spina bifida occulta and exists as a diffuse soft mass of fat, usually over the sacrum.

(2) *Abnormal hair in the lumbo-sacral region.* Coarse hair several inches in length may be found which is maximal in the midline and exists from the time of birth. On occasion a silky down limited to a discrete area is found.

(3) *Angioma.* A superficial angioma may be found in or near the midline in the lumbo-sacral region.

(4) *Dermal dimples.* Skin depressions near to the midline with fixation of the epithelium to underlying layers may indicate the presence of a spina bifida.

(5) *Dermal sinuses.* Dermal sinuses near the midline may mark the outlet of a continuous fibrous or fistulous tract extending directly into the spinal cord but the cutaneous opening may be minute and may not be seen without a very close inspection of the back. These sinuses must not be confused with the common pilonidal sinus which has no connexion with the subarachnoid space and which is situated in the midline.

Progression and Later Stages

The majority of our cases have presented in the manner described or with the foot deformity already established. In the latter cases the mode of development of the deformity is no longer evident; there is already a pes cavo-varus with mobile clawing of the toes and except that weight is borne on the outer border of the foot no muscle can be seen to be overacting. The leg and foot are short and the arch of the apparently unaffected foot is sometimes a little high but within normal limits. When the

foot deformity is established, the reflexes become abnormal and there may be sensory changes. With continued progression, sensation becomes increasingly impaired, trophic ulceration occurs and the deficient circulation becomes more obvious.

Another method of presentation is that of Case 2 (see Table) where the appearances of the lower limbs were similar to those seen in cases of myelocoele. We have not otherwise seen calcaneo-valgus deformity developing after birth, but it is likely to occur probably where the spinal cord is affected at a high level and these cases are more likely to develop paraplegia. Other authors have mentioned such cases with paraplegia or weakness of the lower limbs and an unsteady gait.

A history of incontinence is unusual in early cases but bowel and bladder symptoms may develop later. There may be incontinence which cannot be diagnosed in a young child only just old enough to be expected to learn control (Case 2). Urinary incontinence of the type associated with a neurological bladder which empties itself only when fully distended is difficult to identify. Double incontinence with loss of bladder and bowel sensation may supervene in time.

In this description of the progression of lower limb abnormality it is clear that the syndrome is very similar to that occurring with spinal tumours except that the rate of progress in the latter is very much faster so that foot deformities have not time to develop. Paraplegia is likely to be the end result in both types of untreated case, although rare following spinal cord lesions associated with spinal dysraphism. The foot deformity which develops gradually over a long period in the dysraphism cases precedes the paraplegia.

Our experience of adult cases is limited and it appears that they are not often diagnosed. Their deformity has presumably been static for a number of years and they are unlikely to come for orthopaedic attention except for supply of surgical footwear. Those who develop trophic ulceration may be recognized if the clinician is aware of the syndrome, otherwise they are most likely to come to notice if they develop urinary or bowel dysfunction (Case X). Our personal adult cases were recognized by reason of attendance for supply of footwear, one of them having trophic ulceration.

Radiological Investigations

Apart from clinical examination and periodic review, the other chief investigation required is radiology. Every child (in the absence of a myelocoele) who shows a peculiarity of gait, poor posture associated with a short leg, a short and thinner leg,

progressive deformity of a foot, absent deep tendon reflexes or evidence of sensory impairment in the lower extremities, should have careful radiographic examination of the spine. Lichtenstein (1940) has pointed out that bone anomalies may be absent but it is our experience that plain films usually show congenital anomalies of the vertebral column such as spina bifida, which is the commonest abnormality, or hemivertebrae. A bone septum (diastematomyelia) may be shown. Spina bifida of even the slightest degree is almost always present in this syndrome but the bone abnormality is not necessarily at the site of the lesion, which may be situated some distance away as in Case 2.

Myelography is valuable for more exact diagnosis and for localization of the lesion but it should not be done lightly; its purpose is confirmatory. It will demonstrate space occupying lesions and obstruction to the flow of the opaque medium and if there is sufficiently clear definition of the filum terminale and nerve roots a myelogram may indicate the site of the conus medullaris. Occasionally the spinal cord may be seen to be held against the neural arches instead of being in its normal position lying against the vertebral bodies. Some traction lesions, however, are not demonstrated by myelography. Whilst screening it is necessary to examine the patient supine as well as in the classical positions. We have used myodil as the opaque medium; at lumbar puncture for its injection, cerebrospinal fluid is removed for examination and is usually found to be normal.

Diagnosis

At present the early case is not easy to diagnose confidently and usually the progress of deterioration has to be watched over a long period before the diagnosis can be established. There is a wide differential diagnosis and any history elicited from the parents must be considered with caution. A previous diagnosis of poliomyelitis may be suspect since it may have been applied *post hoc* to explain abnormality. A story of old injury likewise must not be accepted without further enquiry.

In the first place, the diagnosis depends upon the presenting symptoms. The peculiar gait suggests many conditions, a plantar wart, early hallux rigidus, injury to sesamoids underlying the first metatarsal head, foot strain, or too small a shoe. Pain, if present, may bring to mind March fracture, early hallux rigidus, trauma, foot strain, osteochondritis of a metatarsal head or of the navicular, or lumbar nerve root irritation.

The cases of short foot and leg of the variety for which no cause is found are distinguishable from

those due to spinal dysraphism in the course of long term review by the foot shape, function and the gait, which continue to be normal. Other possible causes of a short foot and leg are old bone infection, previous fracture, poliomyelitis and hemihypertrophy or angiomatosis of the other limb.

Disease of the nervous system must be excluded, e.g. neurofibromatosis, tumour of the cauda equina, Friedreich's ataxia, the distal (Gower's) type of myopathy, peroneal muscular atrophy, polyneuritis and cerebral palsy.

In the late case, the limb deformity and the neurological deficit make the diagnosis straightforward.

Treatment and Results

Excluding cases of diastematomyelia with a bone septum evident on a plain x-ray film that ought to be operated on at once, the only treatment possible in the early stages is a passive correction of the shortening by the use of raising on the shoe, with or without irons. This treatment is rarely effective for any length of time. When the presence of a neurological deficit has been established, laminectomy should be performed after myelography. Surgical exploration of the vertebral canal, including opening the dura to permit inspection of the spinal cord, is intended to remove the cause of the disability by releasing the anchoring of the spinal cord or by excising the cause of pressure. The results expected are not that the lower limbs will return to normal, nor that previous incontinence will be cured, although these things may happen if the case is dealt with early enough, but that there will be no further deterioration nor increase in the lower limb abnormalities and deformities. The changes which have taken place in the spinal cord and nerve roots may be irreversible so that recovery cannot occur. The operation is essentially preventive. Orthopaedic treatment will be necessary to correct the established foot deformity and residual muscle weakness. After removal of the primary cause of muscle imbalance, full use can be made of normal function and growth in the correction of any postural foot deformity. Without removal of the primary cause, orthopaedic treatment may temporarily correct a postural deformity but is ineffectual except as a short term expedient.

Illustrative Cases

In the Table we present a summary of our first 16 fully investigated cases. There are many other cases under review some of which are awaiting myelography. The cases in the summary are given in order of myelography and some, which are not cases of the syndrome described, have been in-

Case	Sex	Date of Birth	Date First Seen	Syndrome	Neurology†			Sensation
					K.J.	A.J.	Plantars	
1	F	18.12.46	17.12.56	Poor circulation lower limbs; right foot slightly smaller than left	+++	++	? F	?
2	M	19.5.53	12.1.55	Dragging inverted left foot and everted right foot; progressive paralysis of lower limbs	+ -	- -	E E	Abnormal
3	F	5.3.45	7.9.55	Left pes cavus with clawing and dropping of forefoot; left leg short $\frac{1}{2}$ in.; right foot normal	++	++	F -	Normal
4	F	8.8.45	12.2.57	Inverting left foot only when walking, distortion of shoe, progressive cavo-varus; right foot normal; legs equal length	± ±	- -	F F	Normal
5	F	11.11.46	23.7.47	Originally clawed outer toes left and severe valgus both feet; not seen 1951-55 and then left foot smaller than right; progress: left cavo-varus and clawing; poor skin circulation; sacral naevus and excess hair	++	+	F -	?
6	M	21.12.46	9.6.55	Left cavo-varus; left foot shorter than right; left leg $\frac{1}{2}$ in. short; sacral hairy patch	++	++	F E	Normal
7	F	30.8.50	1.11.54	Right foot smaller than left since birth; developed mild cavo-varus; sacral hair	- +	- +	E F	Normal
8	M	21.8.52	4.11.57	Short left leg and foot; pes cavus R. and L. worse with progressive increase of varus in left; increasing weakness of left foot; falling over frequently	++	++++	? E	?
9	F	18.11.47	17.2.58	Mild pes cavus at birth; developed pain left foot with increasing cavo varus and hypersensitivity and impairment of joint sense; left foot smaller than right; legs equal length; weakness dorsiflexors left foot	++	+++	F F	Abnormal
10	F	30.8.45	14.11.51	Left club foot treated at birth; short right foot and leg developed and increasing cavo-varus of both feet, later left leg was shorter	- -	- -	- -	Trophic ulcer loss of sensation in both feet
11	M	2.2.56	9.2.56	Bilateral club feet with full correction obtained by age of 4 mths; persistent overaction of Tib. Ant. with progressive increase in cavo-varus deformity starting at age of 18 mths; lower limbs becoming a little spastic	+++	+++	E E	?
12	M	17.4.50	7.9.50	Left leg and foot smaller than right from birth; left foot originally very valgoid but aet. 5 yrs started developing cavo-varus; sacral hair	+ -	+ -	F ?	Abnormal
13*	F	26.6.46	1.11.54	Since infancy small right foot; sacral fat pad; developed right claw toes and cavus with increasing cavo-varus; left foot mild cavus	- +	- +	- -	Abnormal
14	M	13.10.54	13.5.58	Persistent inversion left foot; left foot and leg short; pes cavus developing in left foot	++	+	F E	?
15	F	11.12.46	8.3.55	Rapidly increasing left pes cavo-varus; short left foot; legs equal length	++	+	F E	?
16	F	5.7.48	15.7.58	Short right foot and leg, with early right pes cavus; patch of hair over lower thoracic and lumbar region	- +	- +	? F	Abnormal

* Case referred to us by Mr. J. K. Stanger and Dr. Christine Cooper.

† Neurology: ++ = present - = absent

CASES OF SPINAL DYSRAPHISM

Survey Radiograph		Myelography	Comment
Bone septum LV.2; spina bifida LV.3-SV.1	30.3.57	Diastematomyelia	<i>Laminectomy</i> (4.4.57) Bone septum at LV.2/3 level excised; diastematomyelia 1.9 cm.; conus at lower border LV.4
Minimal spina bifida LV.4 and 5 and SV.1; increasing interpedicular widening at LV.1 indicating enlarging tumour	6.11.57	Total block preventing flow to LV.1	<i>Laminectomy</i> (11.11.57) Transverse pressure band from pedicle to pedicle at LV.1; spinal cord was markedly enlarged at and below this level, probably the result of gliosis
No bone abnormality	12.11.57	No abnormality demonstrated	Deformity not progressing after tendon transplants in foot; right foot normal
Multiple anomalies of whole spine; tomograph: narrowing of vertebral canal antero-posteriorly by laminae at LV.5	12.12.57	Filling defect left side of LV.5 level	Parents refused operation; left foot deformity slowly developing
Marked spina bifida LV.2—sacrum	31.1.58	Enlarged filum terminale and abnormally caudal conus	<i>Laminectomy</i> (31.3.58) Traction band at LV.3 extra and intra-dural attaching to filum terminale and left S.1 nerve root and holding spinal cord posteriorly; conus at caudal border of LV.3
Spina bifida LV.5-S.1 with anomalies of vertebral bodies	25.4.48	No abnormality demonstrated	<i>Laminectomy</i> (9.6.58) Traction band, extradural, at LV.5; absence of extradural fat in this area and adherence of ligamentum subflavum to dura; conus at caudal border of LV.3
Spina bifida LV.4 and S.1 (only four lumbar vertebrae)	30.4.48	Diastematomyelia; conus abnormally caudal	<i>Laminectomy</i> (16.6.58) Fibrocartilaginous septum LV.4; ligamentum subflavum adherent to dura in this area with absence of extradural fat; diastematomyelia limited to area of septum
Minor spina bifida at LV.5 and greater degree at SV.1	14.5.58	No abnormality demonstrated; some extra-theal myodil	Undiagnosed
Moderate spina bifida SV.1	23.6.58	No abnormality demonstrated	Physical signs improving; undiagnosed
Marked spina bifida TV.9—sacrum	16.7.58	Diastematomyelia; conus situated abnormally caudal	<i>Laminectomy</i> (22.9.58) Bone septum LV.2; conus not seen, but situated below mid LV.4
Mild spina bifida LV.5 and more severe at SV.1	3.9.58	No definite abnormality demonstrated but theca lay away from the vertebral bodies	Probably cerebral palsy but undiagnosed; not club foot relapsing in the usual manner
Extensive spina bifida of whole lumbar spine and sacrum	19.9.58	No definite abnormality demonstrated	Under review; foot deformity worse; awaiting laminectomy
Spina bifida and gross anomalies mid and lower lumbar, with some lower lumbar scoliosis	6.10.58	Diastematomyelia (minute); conus abnormally caudal	<i>Laminectomy</i> (9.10.58) LV.3 level, intradural band holding spinal cord posteriorly and rotating it; the spinal cord was bifid immediately cranial and caudal to the attachment of the traction band; the conus lay distal to the level of LV.3 caudal margin
Spina bifida of LV.5 with caudal extension of right lamina of LV.4	12.11.58	Appearances not understood; conus situated abnormally caudal	<i>Laminectomy</i> (24.11.58) Traction band LV.5 lamina to dura and transverse pressure band at LV.4-5 level; conus tip at lower margin of body of LV.4
Marked spina bifida LV.3 to SV.1	3.12.58	No abnormality demonstrated; conus abnormally caudal	<i>Laminectomy</i> (25.5.59) Transverse pressure bands at LV.5; absence of extradural fat in this area; conus not seen, probably at caudal border of LV.3
Wide spina bifida lower thoracic, upper lumbar and ? LV.5; conus at about LV.3 level; tomograph: bone septum at TV.9	28.1.59	Diastematomyelia	<i>Laminectomy</i> (2.2.59) Diastematomyelia extending about 7 cm.; the cords were adherent to the bone septum

± = diminished ++ = exaggerated.

Plantar responses: E = Extensor F = Flexor ? = Equivocal — = Absent.

cluded because we thought at the time that the lower limb abnormalities might possibly be ascribed to a lesion of the spinal cord and also because they demonstrate some of the difficulties of differential diagnosis. No fully investigated cases have been excluded.

All of the 10 cases submitted to laminectomy have had demonstrable lesions at operation although the mechanism by which the lesion produced its effect is not clear in every case. In none of these cases has there been any further deterioration and most have improved in an encouraging way. Myelography demonstrated diastematomyelia in six cases and a total block to flow of myodil in one. In the three other cases myelography showed no abnormality in one and in the other two the termination of the spinal cord could be seen to be abnormally caudal.

Four cases are described below in detail, three from our personal series, of which Case 5 had a traction lesion, Case 2 a pressure lesion and Case 14 a combined traction and pressure lesion. Case X (an adult) is given as an example of spinal dysraphism presenting with urinary symptoms resulting from a pressure lesion. This patient was not one of our personal series. He was seen in consultation before operation and is reported here with permission of Mr. Keith Yeates, Dr. John Walton and Mr. John Hankinson.

Case 1 of the series has been reported and discussed elsewhere (James and Lassman, 1958).

Case 5. Female, aged 11 years. This child was first seen at an orthopaedic clinic in March, 1947, when she was 4 months old because the left foot was smaller than the right and the fourth and fifth toes were flexed. The deformity of the toes was treated by strapping and manipulations. In September, 1950, the toes were operated on to correct the deformity. She was not seen between June, 1951 and December, 1955, when she came under our care because she was distorting her footwear. Her left foot was much shorter than the right and there was a tendency to inversion and equinus with dropping of the forefoot. She had a sore on the back of the tendo-achilles and an unpleasant callous under the fifth metatarsal. By July, 1957, the foot condition had altered; the right foot was normal, the left foot was very short, rather high arched with hyperextension activity of the great toe. The muscles of the ankle, foot and toes were functioning, but in voluntary use the invertors overacted. There was still a callous under the fifth metatarsal head. The skin of the left calf was cyanotic and became purple in cold weather and the circulation in the foot was poor. The left leg was at least $\frac{1}{2}$ in. shorter than the right. The left ankle jerk was absent. A pigmented patch to the left side of the sacrum and excessive hair in the lumbo-sacral region were found on examination.

Radiographs of the lumbar and sacral spine demonstrated a marked spina bifida in the lumbar region from LV.2 downwards and also in the upper sacral region (Fig. 2).

Myelogram outlined a midline structure in the terminal theca which was wider in diameter than one would expect for the filum terminale (Fig. 3). This was thought to be possibly anchoring the cord as the conus medullaris lay at an abnormally low level. With this evidence and the clinical history and findings, spinal exploration was considered to be justified.

At operation when the laminae had been superficially exposed, a fatty fibrous mass was found between the laminae of LV.4. This mass was isolated by laminectomy of LV.3 and could be easily lifted off the dura to which it was attached by a fibrous band. The band, accompanied by an artery, passed from the deep surface of the fatty fibrous tissue to the dura, continuing from the deep surface of the dura to the very large filum terminale and to the left first sacral nerve root within the dura. It was approximately $\frac{1}{2}$ in. in diameter and $1\frac{1}{2}$ in. in length and was holding the cord towards the neural arch and away from the vertebral bodies (a state not evident in the myelogram). The spinal cord fell to normal position when the band was divided, but was not seen to shift cranially with the release of tension. The conus lay at the level of the caudal border of LV.3.

The post-operative course was uneventful.

When seen 10 months after operation, the circulation in the left leg was normal and the blue and purple discolouration of the calf no longer occurred. The reflexes

FIG. 2.—Case 5.—Antero-posterior survey view of the lumbo-sacral spine showing marked spina bifida in the lumbar region from LV.2 downwards and also in the upper sacral region.

FIG. 3.—Case 5. Myelography demonstrates in the terminal theca a midline structure which is wider in diameter than would be expected for the filum terminale. The appearances suggest that the spinal cord is anchored down and that the conus medullaris lies at an abnormally low level.

were unchanged. The shortening of the left leg was a little more than 1 in. There was subjective sensory improvement in the foot, in that she could feel, when she put her foot to the ground, a sensory deficit of which she had been unaware before operation. There was increased muscle power and the foot was used in a plantigrade manner, no longer being inverted when walking.

Case 2. Male, aged 4 years. This boy was referred to an orthopaedic clinic in January, 1955, at the age of 20 months because he was not walking on his own. He was healthy and was a full-term baby without neonatal complications. On examination, when he attempted to walk holding on to the furniture, he inverted his left foot and everted his right. When in a standing position, the left foot showed a voluntary tendency to adduction and inversion of the forefoot which the mother had first noticed two months previously. Until then, she had thought that the boy's feet were quite normal. He was not obviously spastic in the left lower limb, but his gait suggested this possibility. He was given irons to wear and slowly gained in strength to be able to walk on his own. Sensation in the legs seemed deficient at about 2 years of age; his lower limbs at this time resembled those of a child with a myelocoele.

He was in hospital for 10 weeks from February to April, 1956, owing to trophic ulceration of the right third, fourth and fifth toes possibly induced by a hot water bottle. During this period, whilst the toes

healed very slowly the deformities of his feet became more severe and more fixed in spite of splintage. By April, 1957, his lower limb power was deteriorating rapidly and he was hardly able to walk barefoot. On October 31, 1957, he was admitted to hospital for investigation, by which time he was unable to walk without assistance even with short irons and boots. He had very little voluntary power of the knees and feet but quite good power of hip movement. It was difficult to determine his degree of sensory loss but he had analgesia to pin prick on the back of the thighs and in the sacral region. The left knee jerk and both ankle jerks were absent and the right knee jerk was just present; both plantar responses were extensor. He had always been doubly incontinent, having no idea when his bladder or rectum were full.

Radiograph of the dorsal, lumbar and sacral spine showed spina bifida of LV.4, LV.5 and upper sacral region with widening of the interpedicular distances with flattening of the pedicles most marked at the level of LV.1 but extending from TV.11 to LV.2 in a lesser degree. The lateral view showed that the posterior aspects of the bodies of these vertebrae were concave. The flattening of the pedicles suggested to the radiologist that the lesion was an expanding one, but he did not think it excluded congenital origin.

Myelography was performed and the myodil flowed upwards as far as the level of the body of LV.1 where a complete block to the upward flow was demonstrated. The radiologist was unable to detect any negative shadow of the spinal cord but several half shadows were present in the myodil suggesting either a gross anomaly or a gross displacement of the cord, probably the former. The myodil passed laterally on both sides into the exit foramen between LV.1 and LV.2 in a manner which suggested a congenital anomaly. The upper border of the myodil in the head down position showed no typical shape but rather an irregular contour in the lateral view.

At operation on November 11, 1957, the spinal cord from TV.12 level caudally to just proximal to the conus was enlarged and white in colour; an extradural fibrocartilaginous band compressed it transversely at the level of the pedicles of LV.1. The cord pulsated cranial to this band and at the conus but not in between, and the band was clearly restricting the flow of cerebrospinal fluid from above downwards. After removal of the band the cord beneath began to pulsate and there was a clear flow of the cerebrospinal fluid downwards. The enlargement of the spinal cord appeared to be due to a congenital abnormality and was not in any way cystic. Microscopy revealed the band to be composed of dense acellular fibrous tissue structurally resembling a ligament.

After operation the boy developed a cerebrospinal fluid fistula and continued to drain cerebrospinal fluid in very large quantities despite further attempts to close the gap. Ventricular drainage became necessary to allow the back to heal and the quantity of fluid discharged through the drain, together with the difficulty of controlling the flow from the fistula, suggested some abnormality of cerebrospinal fluid production. There was no other evidence, internal or external, to suggest

that there was an associated Arnold-Chiari malformation.

After discharge from hospital on January 30, 1958, the strength of his lower limbs developed quite rapidly to begin with but he could not walk without assistance for nine months after operation. Subsequently he began to walk without sticks, wearing a caliper on the right leg. Fourteen months after operation the foot deformities were unchanged; he still lacked voluntary power in the evertors of the left foot and in the invertors of the right, but there has been some recovery in the right calf muscles. Lower limb sensation was difficult to assess but five months after operation his right plantar response was flexor and 12 months after operation a flexor response was obtained on the left side.

His mother stated that approximately one month after discharge from hospital, the boy, for the first time, began to know when his bowels were going to work and after this time he rarely dirtied himself, provided he was near home. As regards micturition, she thought the boy had some feeling but since he wore either a urinal or a napkin he made no effort to control himself.

Case 14. Male, aged 4 years. He was referred to an orthopaedic clinic at the age of 3 years 6 months, because he had been walking with his left foot inverted since the age of 18 months. This tendency was evident in the clinic but there was no real abnormality of the foot apart from slight shortening as compared with the right foot. Examination showed a slight (less than $\frac{1}{2}$ in.) shortening of the left leg; normal right leg reflexes, an absent left ankle jerk and a left extensor plantar response. The left lower limb felt spastic on passive movement.

During the next five months the left foot rapidly developed cavo-varus. The back appeared normal. There was no evidence of bladder or bowel disturbance and the reflexes were unaltered. Sensation was not tested.

Radiographs showed spina bifida of LV.5 and the laminae of LV.4 extending caudally made a curious apparently curved bony ridge crossing the spinal canal at the level of LV.4/5. The interpedicular distance at the lumbo-sacral level was increased (Fig. 4).

Myelography showed no definite abnormality, but suggested that the conus medullaris lay almost at the level of the lumbo-sacral articulation. There were some linear translucencies which could not be satisfactorily explained.

Exploration was decided upon, in view of the neurological deficit in the left lower limb, the rapid development of cavus in the left foot, and the curious myelographic appearances.

At operation the spinous process of LV.4 was found to be expanded on the right side and the right lamina of LV.4 was continuous with that of LV.5. At the level of the pedicles of LV.4 an extradural transverse band firmly adherent to the dura was constricting the spinal cord. Immediately caudal to this transverse band and partly attached to its caudal edge was a cylindrical band 1.0 cm. long with five webbed digital prolongations, which were firmly fixed at their anterior ends to the dura and at their posterior ends to the deep surface of the fatty fibrous tissue which filled the gap. This

FIG. 4.—Case 14. Antero-posterior survey view of the lumbo-sacral spine. The laminae of LV.4 extending caudally exhibit a curious apparently curved bony ridge crossing the spinal canal at LV.4/LV.5 level.

traction band ran obliquely from the midline of the dura in a caudal direction towards the right side in such a way that the whole spinal cord was probably held posteriorly to the right. The traction band was excised, and the transverse band was divided when the dura was opened to expose an apparently normal spinal cord with the conus lying caudal to the transverse band at the level approximately of the intervertebral disc between LV.4 and LV.5. The dura was closed without difficulty but for two or three days post-operatively there was a fluid fistula which healed without further interference. Microscopy of the traction band showed very dense fibrous tissue.

Three months after operation there was no change in the physical signs. At six months, the reflexes were unaltered and there had been no change in the degree of cavo-varus as compared with the state before operation; progression had ceased.

Case X. Male, aged 26. Up to the age of 11 years he had suffered incontinence of faeces and when aged 18 years a small swelling had been removed from over the coccyx. He had begun to experience increasing difficulty in passing urine for two years before his admission to hospital for investigation. Whilst in hospital and shortly after a myelogram, he developed acute retention of urine.

Examination showed sensory loss around the anus and perineal region and on the buttocks in the distribution of the fourth sacral and first coccygeal segments;

there was no abnormality in the lower limbs. Survey radiographs revealed spina bifida of the lower sacral segments S3-S5 which was remarkably wide at S4 and S5.

At myelography, the myodil flowed downwards to the level of the S3/S4 segments. In the supine position the myodil column divided into two lateral columns at the level of the lower border of S5, leaving an oval shaped defect in the outline of the myodil. In the lower lumbar and upper sacral region the appearance of the myodil column suggested that the spinal cord was extending to a very low level.

At operation Mr. Hankinson found a lipoma attached to the skin in the region of the sacral bone defect. The lipoma passed through into the vertebral canal and was continuous with the dural sac and within the dura it was firmly attached to the spinal cord. The spinal cord was of normal thickness and extended to the lowermost portion of the sacrum; the spinal nerve roots passed horizontally to escape through their respective intervertebral foramina in the manner usually seen only in the early stages of human foetal development. The attachment of the lipoma at the periphery of the bone defect was freed and the dura left open and covered with gelfoam. Spontaneous micturition slowly returned and two weeks after operation catheterization was no longer required. The difficulty with micturition has continued to decrease.

Discussion

The picture of this syndrome and its causative lesions which we have endeavoured to draw is based on information from the literature and on a personal series of 16 cases completely investigated and submitted to myelography and more than 20 clinical cases under review and as yet unproven. All these are children. The unproven cases do not include the many cases seen which for no known cause have one leg shorter than the other and a small foot, a combination which is remarkably common in children's orthopaedic practice. Many cases produce a foot deformity which does not progress and, not having developed a neurological deficit, do not appear to justify full investigation and laminectomy. Some of these cases no doubt would have a lesion demonstrable on myelography and surgical correction of the lesion would prevent the full development of deformity but since myelography is dependent on the introduction of a foreign substance into the subarachnoid space it is not an investigation to be carried out without considerable forethought and good reason. Indeed, our policy has been to undertake myelography only if we expect to proceed to operation when the myodil will escape. We have used myodil in our cases and it has been the material used as a routine by the Department of Neuro-radiology for a number of years. So far there has been no complication occurring from its use

and as we gain further experience with myelography we may be able to change our criteria and use it more frequently. The technique of myelography and the understanding of the radiograph appearances need expert management and we are most grateful to our colleague, Dr. Gordon Gryspeerdt, for his skill in this technique and his guidance in interpretation.

It is becoming apparent from our operated cases that some of the traction lesions may give little or no evidence of their presence by myelography. Case 5, reported above, is an example. The myodil column must be examined throughout the whole theca from the cervical region down to the sacrum; Shorey (1955) has reported a case with lesions at two levels, thoracic and lumbar.

If a myelogram fails to demonstrate a lesion which is in fact present, it fails also to provide a localization of the site to be approached surgically. A normal myelogram thus deprives the surgeon of a valuable piece of information since the lesion affecting the spinal cord may not lie in the immediate vicinity of the bone defect. In some of our cases with a normal myelogram we have been able to operate and find a lesion only because the pattern of spina bifida resembled that occurring in a case previously explored and thereby suggested the likely site and the type of lesion to be anticipated.

All our investigated cases except one have spina bifida occulta, varying in degree from the barely noticeable to the very extensive. The one case with no bone anomaly shown on a radiograph had a normal myelogram; the short leg and pes cavovarus show no further deterioration in the last 12 months and there is no neurological deficit. This child has not been operated on.

As we have already pointed out (James and Lassman, 1958), there is no published information about the normal level in the vertebral canal of the conus medullaris during the years between birth and full growth although we have post mortem evidence suggesting that the ascent of the spinal cord is normally completed by the end of the fifth year of life. Where myelography can demonstrate the level of the conus and when this can be regarded as abnormally caudal, there can also be assumed to be a traction lesion which should be dealt with surgically. Many of these traction lesions will show no other abnormality on myelography and they are the most difficult to diagnose and to localize. Tomography can be of assistance in some cases, notably those with a bone septum in diastematomyelia and those with inversion of laminae where the demonstration of the size and locality of bone anomaly assists the surgeon in the planning of the

operation which must, of course, involve as few laminae as possible. Only two of our cases needed excision of four pairs of laminae; the remaining eight had three pairs removed.

Pain has not been a factor in the symptomatology of our cases, nor has the spasticity reported by other writers, and this is possibly because we have been able to see our cases at an earlier stage in their history. Neither has ataxia in the form of clumsiness and unsteadiness been evident (except in Case 2), but it is obviously likely to occur at a later stage. Low back pain or stiffness occurring on getting up in the morning and passing off with activity has been referred to by other authors but has not been complained of in our series. The two most important early clues to indicate a possible lesion affecting the spinal cord or cauda equina are either a short leg and foot or a circulatory defect of the skin of the lower limb (as in Cases 5 and 2) of the type seen following poliomyelitis and associated with spina bifida cystica (see also case reported by James and Lassman, 1958).

We wish to stress again the importance of investigating any statement by the parents as to causation of the deformity. Just as other clinicians have done, we have seen many cases where a deformity has had to be considered as due to poliomyelitis because no other cause appeared to be likely; this diagnosis has naturally been passed on to the parents in spite of the absence of any other real evidence of this infection in the past. Injury also has often been advanced by the parents as their own idea of the cause; it is surprising how many children have been dropped in infancy. One adult found to be affected attributed the foot deformity to an injury caused at the age of about 1 year, when her forefoot had caught under a door. She and her family had been satisfied to accept this explanation ever since. Her left foot was normal but the right foot had markedly diminished sensation, pes cavus, clawed atrophic toes and trophic ulceration. Her right leg was $\frac{1}{2}$ in. short. She had two hairy patches on her back and radiographs showed congenital anomalies of the lower lumbar spine and sacrum. The few adult cases we have seen have not been investigated to the extent of myelography. Their deformities, neurological deficits and trophic lesions have been in existence too long to be likely to benefit by laminectomy. They have been treated as conservatively as possible in the same way as cases of spina bifida cystica.

It is clear that the types of abnormality associated with occult spinal cord lesions are similar to those associated with overt spinal cord lesions, although the severity of the physical disability in the latter is

usually much greater. The essential difference between the two types of lesion of the spinal cord is that the occult lesion is usually extrinsic, whereas the overt lesion is an intrinsic lesion of the spinal cord itself. The latter is therefore much more severe and results not only in failure of spinal cord development but also in failure of formation of normal tissues which should be developing synchronously with the spinal cord. The extrinsic lesion occurs slightly later in foetal development; there may be no associated failure in other tissues and its principal effects occur with growth of the child after the associated tissues have been formed; the subsequent abnormalities therefore develop gradually and later in life. As has already been indicated in the syndrome we have described, the established deformity of pes cavo-varus resembles a type of relapsing club foot. If the extrinsic lesion of the spinal cord were to have an effect *in utero*, the pes cavo-varus would possibly have the appearance at birth of a typical club foot because the muscle imbalance would have started earlier and would have been able to advance more rapidly. After birth there are restraints upon this muscle imbalance by the establishment of coordinating reflexes and by the decreasing plasticity of the bones of the foot which provide a mechanical obstruction.

In this communication we have made statements which are based on facts and experience; we have avoided theory as far as possible, but as we continue our clinical and surgical investigations we believe that it may be possible to classify some types of foot deformity as being caused by occult spinal cord lesions. Some cases of club foot are associated with meningomyelocele but the aetiology of other cases is still unknown, and it seems possible that some of the other cases which relapse repeatedly and necessitate local surgery may be due to an occult spinal lesion of the type which we have described here.

The well known association of pes cavus and spina bifida occulta may possibly be now explained but all cases of pes cavus do not necessarily fall into this class and there is obviously need for a reclassification by specifying with greater care the exact form of the pes cavus.

Sufficient information has been obtained to conclude that it is possible to diagnose these cases early and to perform preventive laminectomy without waiting for foot deformity to become fixed. Case 5 demonstrates that with such early procedure muscle balance can become normal or nearly so. Altered bone shape in the older child resulting from abnormal muscle action cannot return to normal but in the very young child almost complete recovery

in this respect may be possible with the return of normal muscle function.

Summary

The purpose of this communication is to emphasize that many types of spinal lesion which have been reported individually are different manifestations of spinal dysraphism and that it is important to recognize their common pattern of symptoms and signs so that an early diagnosis can be made and further deterioration and disability prevented by surgical removal of the cause.

The spinal lesions are classified in three groups, as traction lesions, pressure lesions and combined (traction and pressure) lesions. One case of each type of lesion, proved by operation, is described.

The symptoms and signs facilitating early diagnosis in childhood are discussed. The commonest and earliest evidences are shortening of one foot and leg or circulatory deficiency of the skin of the legs.

The progression of the syndrome by stages to severe neurological deficit, to deformity of the lower extremities and possibly to paraplegia is also described.

The necessity for myelography as a diagnostic procedure is indicated and its uses are discussed.

It is valuable as a preliminary to laminectomy in demonstrating not only the nature of the lesion but also its site. A normal myelogram does not exclude the presence of a lesion. Laminectomy as a method of treatment can be expected only to halt the progressive deterioration. In some cases, however, partial recovery will occur, but such a possibility can only be hoped for and the main purpose of operative treatment is to prevent further disability.

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