The Turkish Journal of Pediatrics

A Quarterly Publication

VOLUME: 7 JANUARY 1965 NUMBER: 1

SPINAL DYSRAPHISM

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Definition

Twenty-five years ago Lichtenstein revived the term *Spinal Dysraphism* to indicate the failure to complete development in the midline of the dorsum of the embryo. This failure may involve cutaneous, mesodermal, neural or entodermal elements, and there is frequently a combination of these.

When we consider the early embryology of the skin of the back, the central nervous system and the spinal column, it is remarkable that mistakes do not occur more frequently. The principle types of disorder with which we are familiar are the various forms of meningocele, diastematomyelia, persistent neurenteric canal, dorsal intestinal fistula, dermoid cysts and sinuses, mediastinal and mesenteric cysts. There are many variations, some so rare that few of us are likely to see more than a small proportion during our professional life.

In 1960 Bentley and Smith published a paper on spinal malformations which offered an explanation of these developmental errors. This paper, which had the sub-title "The Split Notochord Syndrome", makes it far easier in my opinion to understand the maldevelopments which we find, simplifies the matter and is a reminder always to search for multiple lesions.

Briefly the theory is as follows: the notochord appears to be of importance in determining the mesenchymal condensation which leads to the formation of the vertebrae. In some animal embryos which have besen tudied the notochord is known to split occasionally in a sagittal direction, as a single aberration of development. A similar split is believed

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to occur in man. Through this gap the entodermal and endodermal elements of the embryo may come in contact and cells may migrate, or herniate through in either direction. For example cells destined to form skin may pass deeply and later form a dermoid sinus or cyst. This dermoid structure may lie posterior to the spinal cord or in the cord itself or even ventrally in the mediastinum. In the opposite direction gut anlage may pass dorsally, giving rise to the series of anomalies in which enteric tissue is found involving the spinal column or cord. In nearly all cases the width of the spinal canal is increased, which is a reminder of the possible early doubling or splitting of the mesenchymal structures.

The few examples of spinal dysraphism which are now to be described may be said to be of the occult type, that is, the type in which the essential spinal lesion may have very few outward signs but which may in time lead to disablement. These are difficult to place in groups because variation is considerable but I refer mainly to: 1. *diastematomyelia*, i. e. a bifid spinal cord which may have a spur of bone or other tissue between the half cords,

2. *tethering of the cord* by anomalous fibrous bands or a tight filum terminale,

3. lesions which may cause pressure on the cord, for example constricting bands, dermoid cyst, lipoma, hamartoma, anomalies of the bony column, such as enversion of the laminae.

Combinations of these may occur and it will be seen that often there is a lesion present which will interfere with the normal "ascent" of the spinal cord as the child grows. Such effects of traction will become apparent not necessarily at birth but during the first few years of life.

The syndrome of the occult form of spinal dysraphism may therefore include as traction develops:

- a loss of sensation in part of the lower limbs,
- chronic or recurrent ulceration of anesthetic skin of the toe,
- a change in the style of walking, which may be due to muscle weakness or increasing deformity of the foot, or diminished rate of growth of foot or leg on the one side,
- diminished knee or ankle reflexes,
- there may be signs of slow compression within the spinal canal, which in children may be indicated only by backache, back stiffness, constipation or frequency of micturition, long before neurological abnormalities are found.

Commonly there are helpful stigmata on the back which provide clues to the presence of underlying disorders. There may be an area of hair-bearing skin on the back (Figure 1). A pigmented scar my be visible

(Figure 2) or a skin dimple (Figure 3) or an area of telangiectasis (Figure 4).

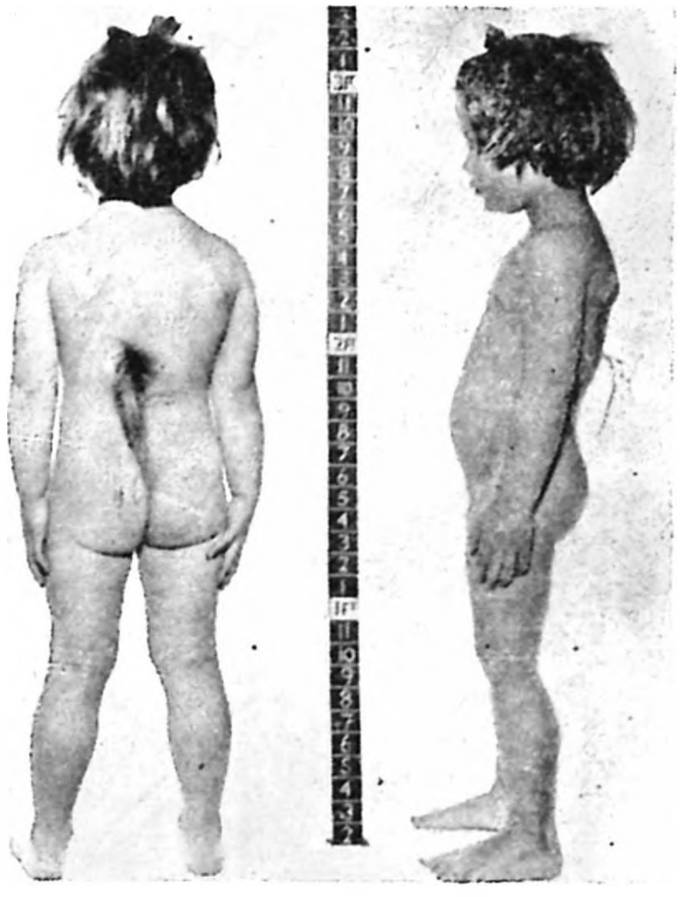

Figure 1.

Figure 2.

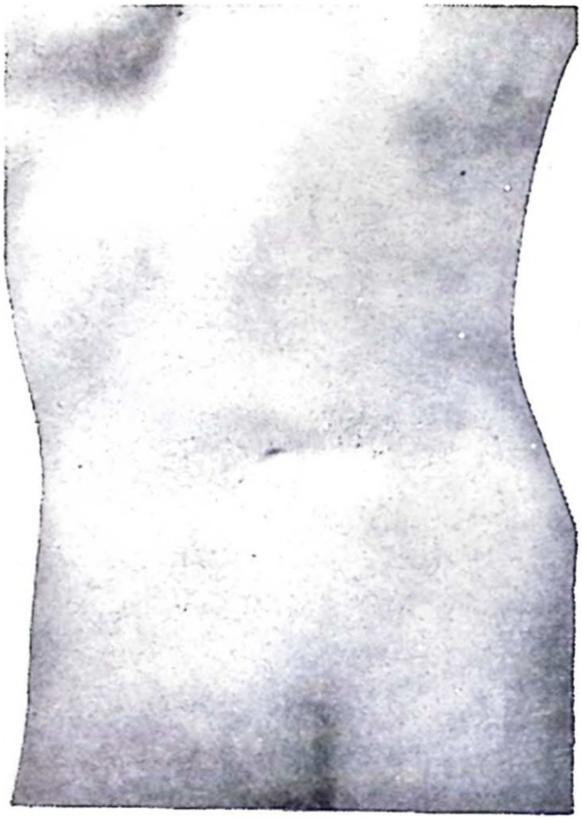

Figure 3.

Figure 4.

Radiology of the spine reveals congenital anomalies, some of which are not in any way the cause of symptoms or signs. Their presence, however, is an indication that a harmful lesion may be found if it is sought. There may be spina bifida, hemivertebrae, abnormal laminae (Figure 5). A bony spur may be apparent (Figures 6). The spur may be extremely

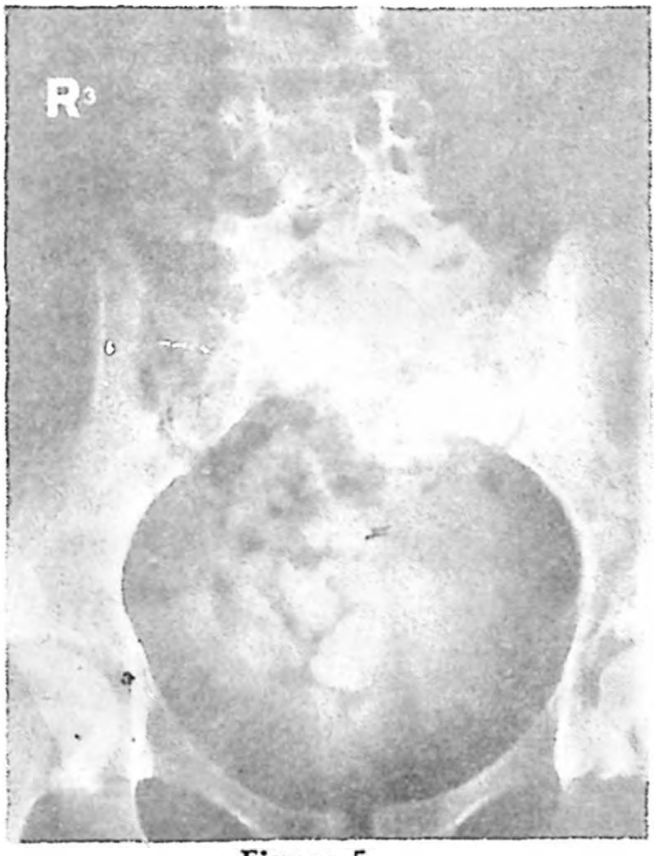

Figure 5.

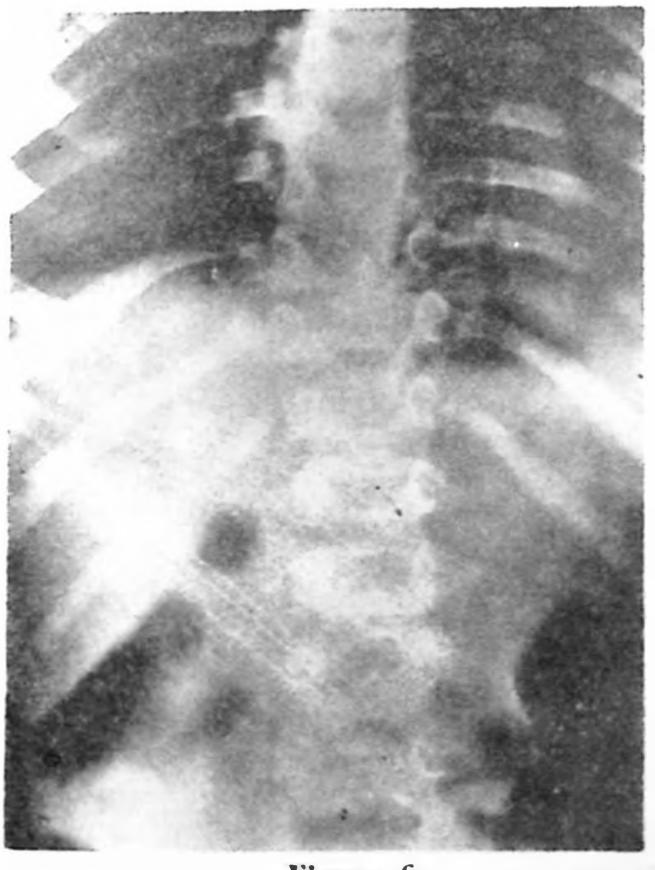

Figure 6.

small and not easily seen. Signs of an expanding lesion within the spinal canal are shown in Figure 6 in which the dermoid cyst lay cranial to the bony spur. The pedicles of the neighboring vertebrae are separated and thinned.

Myelography is required if a more complete diagnosis is to be made and this should be performed by the cisternal route. Gryspeerdt has shown how informative this technique can be. The findings which may be obtained include the following:

1. Space occupying lesion in the neighborhood of a bony spur (Figures 7 and 8).

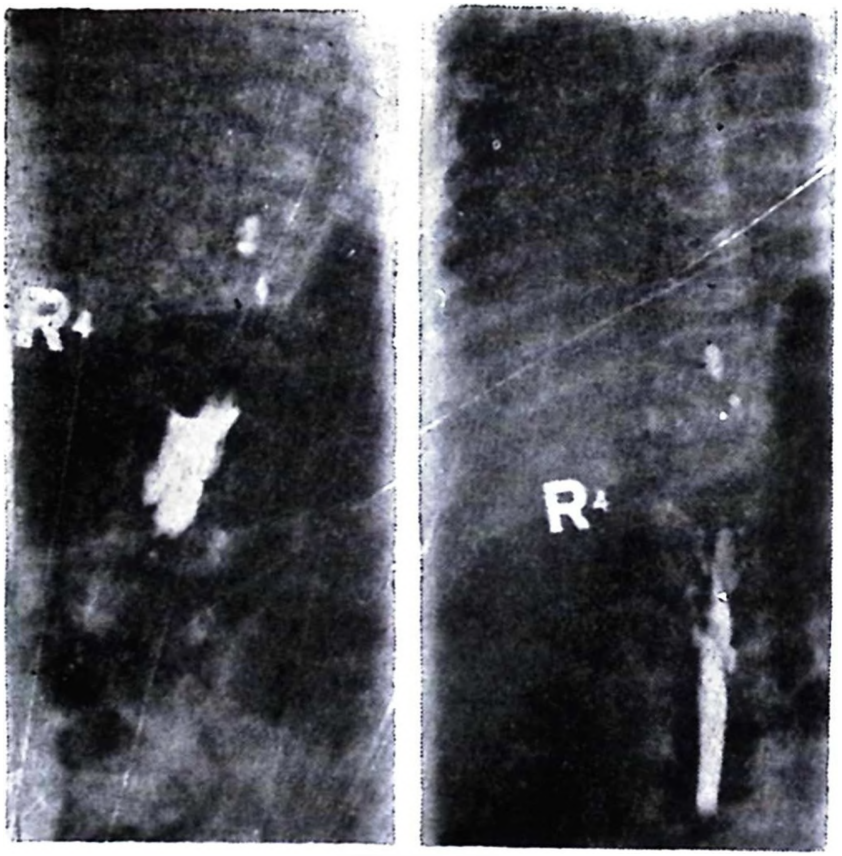

Figure 7.

2. Diastematomyelia indicated by a persistent defect surrounding a bony spur (Figure 9).

3. The level of the conus may be seen to be at an abnormally low level. The conus may be visualized as in Figure 10. The conus is often attenuated and leads to a filum terminale, seen in the same figure, which is often wider than normal. When fluoroscopy is carried out with the child in the prone position the anterior spinal artery may be visualized (Figure 11). When the artery reaches or passes the level of a defect as in this figure, there is clear evidence that the cord lies caudal to the defect and is hence probably suffering from traction or tethering.

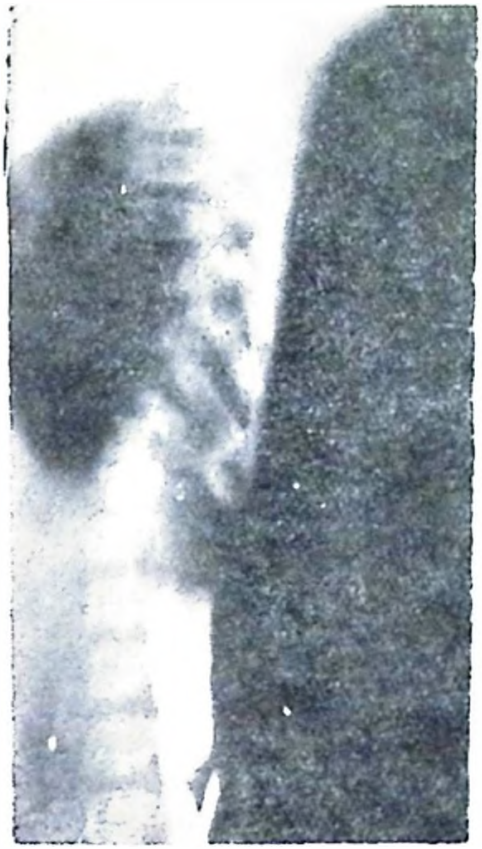

Figure 8.

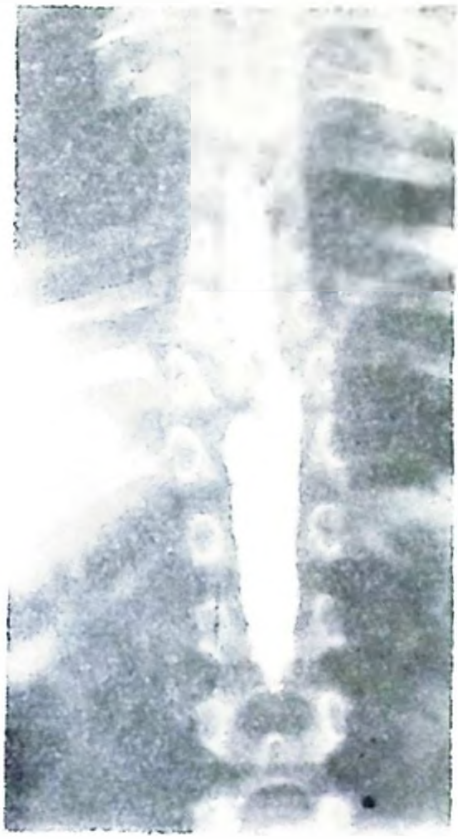

Figure 9.

Figure 10.

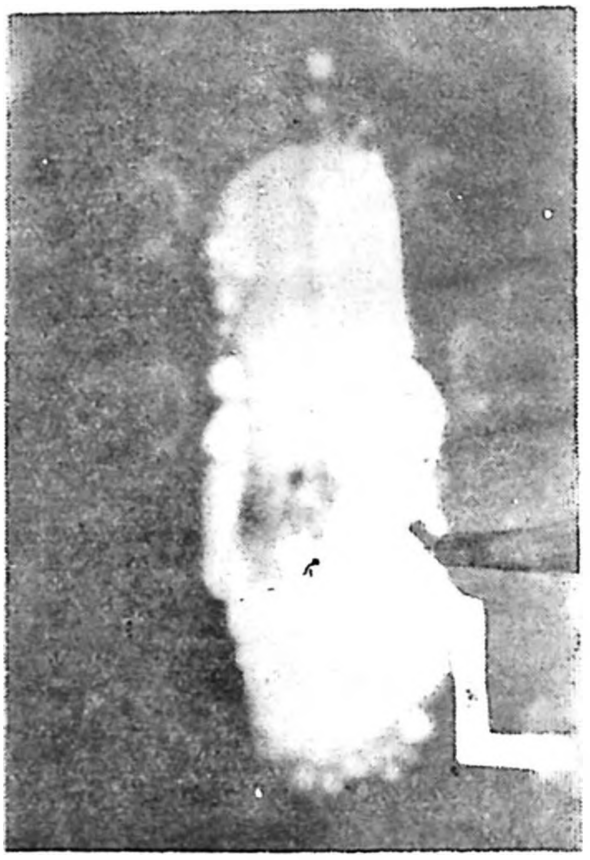

Figure 11.

Findings

It is not possible to describe all the types of anomalies which may be found but Figures 12 and 13 are examples of cord tethering.

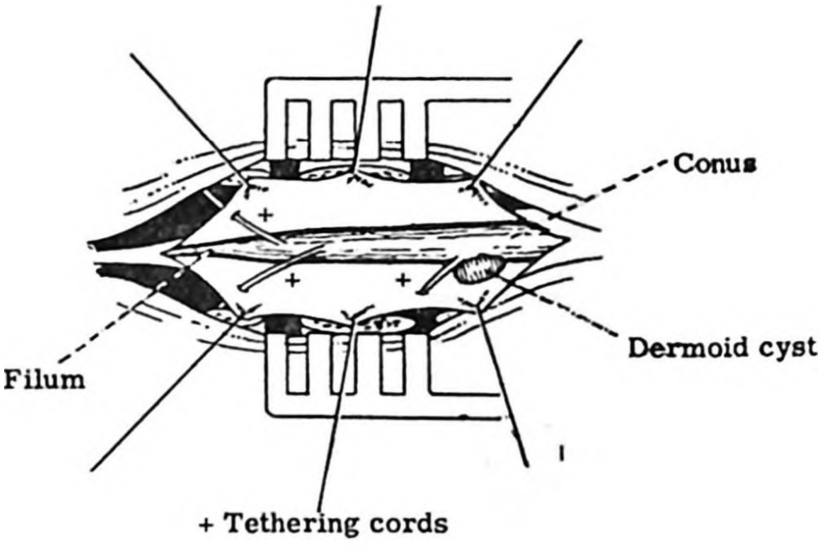

Figure 12.

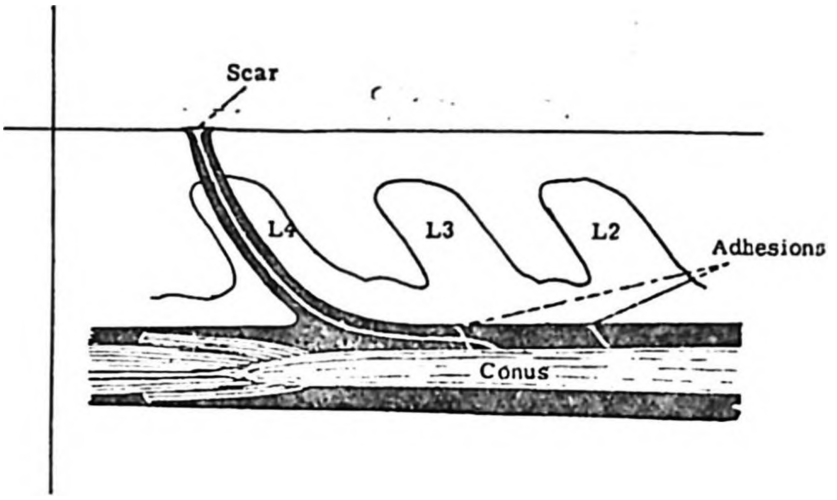

Figure 13.

Conclusions

It is suggested that the indications for further investigations are as follows:

The presence of a peculiar gait, a shortened or thinned leg, unequal feet, deformed foot, absent deep reflexes, sensory loss or chronic skin ulceration, and skin stigmata as described above.

Indication for operation exists when there is a clear history of a progressive disability or when myelography indicates the presence of an abnormally low conus or when there is evidence of a space occupying lesion. Further indications exist when there is a dermal sinus, which may be responsible later for deeper infection.

Proof of benefit to the child in many cases is lacking but if the above indications are used it is reasonable to consider surgical intervention to be of a prophylactic nature.

James and Lassman, to whom much is owed for their studies of these conditions, agree that improvement of function of the lower limbs is often found after operation.

REFERENCES

Bentley, J. F. R., and Smith, J. R.: Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome, Arch. Dis. in Childh. **35:** 76, 1960.

Gryspeerdt, G. L.: Myelographic assessment of occult forms of spinal dysraphism, Acta Radiologica **1:** 702, 1963.

James, C. C., and Lassman, L. P.: Spinal dysraphism, Arch. Dis. Child. **35:** 182, 1960.