Spinal dysraphism in children

Maurice Choux

Spina bifida occulta is one form of spinal dysraphism, a term used to refer to incomplete formation of the midline structure of the dorsum. It is therefore related to all the other forms of spina bifida which have been described under different names: spina bifida aperta or cystica.

The basic differences between spina bifida cystica and spinal bifida occulta result from the nature of their origin. The spina bifida cystica is a failure of development of the spinal cord tissues intrinsically, with added destructive processes in utero, and cellular hyperplasia, as well as brain anomalies. By contrast, in spina bifida occulta there may be some dysplasia of the spinal cord but none of the other destructive processes associated with cystica; Arnold-Chiari malformation is very unusual in spina bifida occulta.

In cystica, clinical abnormalities are present at birth because of the absence of nervous tissue. However, in occulta the anomalies are extrinsic to the spinal cord. Clinical abnormalities usually develop later.

Normally, the nerve tissues change position with movements of the spine, particularly flexion and extension. During a long time it was thought that clinical symptoms developed later because the conus was fixed in low position by extrinsic factors as lipomas, adherences and so on. This theory was based in part on the fact that in most cases of spina bifida occulta the conus medullaris is found under the L 2 level. In our statistic the conus medullaris is situated lower than normal in the vertebral canal in 81% of the 88 cases. We know now that absence of "ascension" of the spinal cord as cause of neurologic deficit is incorrect. Barson, in 1970, examined the normal spinal cord of 252 foetuses and children aged between 13 weeks gestation and puberty. He concluded that the normal spinal cord ends L 3 at 30 weeks, L 2 at 40 weeks, reaching the normal level at L 1 - L 2 about 9 weeks after full-term gestation. We think that the main factors of secondary neurological deficit are repeated traumatism on a rigid, a fixed spinal cord and possibly vascular deficit.

The abnormalities grouped under the term "spinal dysraphism" include:

1. Malformations of the meninges (excluding the posterior meningocele): anterior, lateral, intrasacral meningoceles, Arachnoidal cysts.
2. Malformations of spinal cord and roots: Fibrous bands between cord and dura, abnormal roots, tethered cord, diastematomyelia
3. Lipomas (subcutaneous, extramedullary, intramedullary, in the filum terminale).
4. Dermal sinus, dermoid cysts.
5. Neurenteric cysts.

It is interesting to note that in many patients some of these lesions are found together.

Spinal dysraphism must be separated from the most common myelomeningocele or meningocele which are grouped under the term spinal rachischisis.

I. Clinical presentation

1. Cutaneous abnormalities
   - Hypertrichosis
   - Subcutaneous mass
   - Skin dimple
   - Capillary noevus
   - Congenital scar

2. Orthopedic problems
   - Unequal development of a lower limb.
   - Deformities as talipes, equinovarus, metatarsus varus, pes cavus, fixed flexion of toes, knees, hips.
   - Scoliosis or kyphoscoliosis.

3. Neurological problems
   - Muscle atrophy
   - Difficulty in walking
   - Reflexes modification
   - Leg pain
   - Sensory loss
   - Trophic skin changes
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4. Urinary problems
   - Incontinence
   - Infection

5. Meningitis

   In our initial serie of 63 cases of spinal dysraphism 16 children were hospitalized for urinary problems, 13 for orthopedic problems and 4 for meningitis.

II. Radiographic findings

Plain Radiographs
   - Anomalies of neural arches
   - Widening of spinal canal
   - Bony spur
   - Anomalies of vertebral body:
     - division
     - hemivertebra
     - puzzle
     - fused vertebral bodies
     - narrow disc space

Myelography and C.T. Scan

Spinal C. T. Scan is needed in all the cases, associated or not with a myelography. The extension of the lesion, associated malformation may be appreciated.

We present 138 observations of occult lombo-sacral malformations in children. At the same time were observed 310 cases of spina bifida cystica (myelo-meningocele and posterior meningocoele).

In our series 38% were diagnosed in the first year. A female preponderance is noted (63%).

The lumbar and sacral malformations interesting the meningeal and the neurological elements cannot be classified easily as well in an embryological as in an anatomical point of view In fact they are rarely isolated and more often associated with osseous, visceral and cutaneous abnormalities.

I. Malformations of the meninges (14 cases)

A.— The abnormalities of the lombo-sacral cul de sac are essentially a narrowing, a dilatation and an irregular dura mater cul-de-sac.

B.— Meningoceles
   1.— Anterior and lateral meningocele are very exceptional in lumbar level (4 cases). Anterior sacral meningocele are more frequent and more than 130 cases are published in the literature, especially in adults. The initial symptom can be in pelvic mass, urinary dysfunction, constipation, but neurological signs are rare. X-rays show a sacral defect and myelography visualize a pre-sacral accumulation of liquid but the passage can be narrow.

2.— Intra-sacral meningocele (2 cases) must be distinguished from dural cul de sac dilatation. The clinical manifestations are pain, urinary symptoms, neurological deficit. X-ray show a sacral canal enlarged.

C.— Arachnoidal cyst (6 cases)
   1.— Extra-dural cysts (3 cases) are more frequent at the dorsal level. The cyst is posteriorly situated and communicated with the sub-arachnoid space in half of the cases. The initial symptoms are pain, neurological deficit or kyphoscoliosis (30% of the cases).

2.— Intradural arachnoid cyst (4 cases) are very rare and confusion with cystic arachnoiditis is possible.

3.— Perineural cysts are rare in children.

II. Malformations of spinal cord and roots (34 cases)

The majority of these malformations are described as dysraphic spinal lesions. Abnormalities of roots = 4 cases (size, distribution, number, adherences) are less frequent that abnormalities of the conus.

1.— In the tethered cord syndrom (19 cases) the spinal cord is in low situation and attached to the dura-mater by a short and thick filum terminale. Clinical manifestation are walk disturbances, foot deformities, pain, urinary symptoms. X-rays show a spina bifida in near by all the cases.

2.— The diastematomyelia (16 cases) is the association of a spinal cord duplication and bone abnormalities especially. Symptoms are walk disturbance, spinal deformities, cutaneous symptoms, sphincter signs. The classical triad: cutaneous signs, spinal deformity, abnormalities of the legs is present in the great majority of the cases. X-rays finds always bone malformations (hemivertebra...).

III. Lipomas (67 cases)

Lipomas are situated in the subcutaneous space and the spinal canal (extra dural and/on intraduraly). The
adherences with the conus in low position, raise several problems for the removal of these tumors. Clinical symptoms are cutaneous signs (lumbarmass,dimple) sphincter deficiency and legs abnormalities. Vertebral malformations are present in all the cases. Surgery is useful on the majority of the cases of lipoma, in the way to separate the attached conus and the subcutaneous lipoma.

IV. Dermal cases sinus; fistula, dermoide and epidermoid cyst (16 cases)

They are more frequent in the lumbar and sacral region and can be associated together. The minor category is represented by a cutaneous fistula. The second category is the dermal sinus extended to the vertebral level. In the more severe category the dermal sinus is in the spinal canal. Associations are possible with a cutaneous fistula or an intradural abscess. Clinically a meningitis may be the first symptoms X-ray show a vertebral defect in many cases.

V. Neurenteric cyst (2 cases)

They are rare in the lumbar level. They are characterized by an anterior communication between the arachnoid space and the retroperitoneal space through vertebral abnormalities. We present two cases of neurenteric cyst and the clinical manifestations were a meningitis once and urinary signs in the other case.