Spinal Dysraphism

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Spinal dysraphism encompasses a spectrum of congenital conditions resulting in a defective neural arch through which meninges or neural elements may herniate. These conditions include spina bifida aperta, spina bifida occulta, meningocoele, myelomeningocoele, lipomyelomingeningocoele, myeloschisis, and rachischisis - names given variably according to radiological or pathological findings. These variations can be grouped as open if the overlying skin is not intact, pending leakage of cerebrospinal fluid, and occult if the defect is well covered with full thickness skin. The two groups call for quite different approaches.

Prevalence

Prevalence of spinal dysraphism has been on a decline worldwide in the past 2 decades. Many factors are likely involved. Better nutrition of women, timely folate replacement, better antenatal care with high resolution ultrasound resulting in an available option for termination for mothers diagnosed with such a defective foetus and liberal assay of maternal serum alpha-fetoprotein are all contributing factors. In Tuen Mun Hospital, with an annual delivery of more than 5000, the incidence of open spinal dysraphism is about 0.2/1000 live births.

Embryology

The first 2 months of embryogenesis can be divided into 23 stages. Around day 18 at stage 8, the neural plate is formed, followed by neural folds and their subsequent fusion. Neuropore closure is to follow and completed by stage 12 around day 28. When caudal neuropore fails to close, open dysraphism ensues. From then until day 56, secondary neurulation sets in forming the spinal cord distal to the second sacral vertebra. Defective secondary neurulation results in occult dysraphism in which the caudal part of the spinal cord remains connected with the epidermis by tissues of mesenchymal origin - the ultimate cause for tethering as the patient grows. (Fig. 4)

Symptomatology

For open dysraphism, symptoms are referable to leaking of CSF and an exposed spinal cord. If CSF leakage is not attended properly, meningitis may ensue adding significantly to its morbidity. The extent of sensorimotor deficits would depend on the level of the meningocoele - the higher the worse. It may be high at the cervical level in about 3.9% of spinal dysraphism.9 In severe cases, sphincter dysfunction may present with rectal prolapse. Chiari malformation presentation varies from major brainstem and lower cranial nerve deficits to fairly asymptomatic and insidious.

With the advent of non-invasive investigations and clinical vigilance, diagnosis is generally made early even for occult lesions. At the time of diagnosis, these
patients are usually neurologically intact only to deteriorate later on. Reasons are manifold. As the child grows, disproportionate lengthening between the vertebral column and spinal cord puts the conus under increasing tension precipitating neurological dysfunction.\(^6\) Activities that result in an abrupt flexion of the spine are also implicated as a cause of trauma to the spinal cord.\(^1\) Repeated mechanical shock transmitted via the subcutaneous tissue up the spinal cord, minor as they may be individually, would in the long run pose another cause for neurological deterioration as the patient grows into adulthood.\(^{2,4}\)

Common complaints include pain, sensorimotor deficits and sphincter dysfunction. Pain is usually at the lower back. It may be associated with posture that leads to stretching of the spinal cord or even on prolonged lying down.

In transitional lipoma, probably because of the extensive involvement, motor deficit is more common. Loss of motor neurons results in smaller calves or buttocks. If passed un-noticed, abnormal gait as the child starts to walk and run would alert parents of the problem.

Bladder dysfunction is even subtler for infants. Delayed toilet training is the usual story but tends to be ignored. The author has patients with sphincter problems all their life until the age of 12 when the diagnosis of lipoma was finally made.

Response to treatment differs among the three groups of symptoms. Most rewarding is pain which usually subsides soon after untethering procedures. Motor and gait improvement is also possible but takes time. Chronic bladder and sphincter problems, however, respond poorly to untethering. Long-term urological attention is likely.

**Diagnosis**

Decades ago, an open dysraphic lesion was diagnosed only when the baby was born and it invariably came as a surprise to the obstetrician, neurosurgeon and of course their parents. Nowadays with thorough antenatal care and investigation, diagnosis could be made early in pregnancy with ultrasound. Counselling could be done with parents and for lesions carrying good prognosis - small caudal lesions without other teratogenicity - the pregnancy is advised to continue and surgery then would impose additional difficulties as it would then be a much bigger operation.

For occult lesions, suspicious stigmata along the midline noted after birth would usually lead to further investigations. Radiological finding of bifid spinal laminae due to the intervening tissues gives this group the name spina bifida occulta. MRI is now a standard investigation demonstrating clearly the nature, the level and extent of tethering.

**Management**

Proper management of children with spinal dysraphism calls for a multi-disciplinary approach. Clinicians have to be aware of how subtle the presenting signs and symptoms may be and follow with appropriate investigations and bring them to the attention of neurosurgeons. The pathology demands also the expertise of paediatric urologists as well as orthopaedic surgeons. Physiotherapist and occupational therapists also have an important role in their rehabilitation. They need to be followed up till their late teens to rule out possible retethering.

**Open Dysraphism**

With the possibility of prenatal diagnosis of open dysraphism, attempts have been made in a few centres in the world to close the defect in utero with the belief that associated hydrocephalus and Chiari malformation could be avoided at birth. Success has been reported. Long-term results and benefit of such an approach are still to be seen.\(^3\)

The aim of surgery in these cases is to free the placode from the surrounding abnormal skin and reduce it into the spinal canal which is closed in a watertight fashion. Epidermal tissue has to be meticulously trimmed from the placode to avoid the late complication of dermoid formation. The major early complication of this procedure is wound healing problem with CSF leakage and meningitis. This could be avoided with multiple layer closure or even double-breast closure utilising lumbo sacral fascia on the sides. For huge defect, a proper skin closure may be difficult if not impossible without requiring rotational flap from the sides. Worst of these cases are those with kyphotic deformity of the spine. Expertise of orthopaedic and plastic surgeons may then be essential.

Equally important as the surgery itself is postoperative nursing care. The main wound has to be protected with light gauze and transparent waterproof dressing. It has been a standard protocol for the author to put the patient in the prone position for the first few days until good union is ascertained. In the prone position, excreta will flow forward leaving the dressing and wound unscathed diminishing the possibility of infection.

In unexpected cases in which the cyst is ruptured and CSF already leaking, antibiotic coverage and delayed closure is preferred.

Patients should be monitored with CT for hydrocephalus. Ventricular shunt could be done, usually at a separate setting from closure of the spinal defect.

**Occult Dysraphism**

Whether surgery is indicated for asymptomatic cases has been debatable.\(^4\) But with a better understanding of the pathophysiology and natural course of the disease, the contention is gradually coming to an end. Total or near-total resection of a lipoma while the patient is just a few months old with meticulous microsurgical technique as advocated by Dachling Pang has been shown to lead to a better long-term outcome.\(^7\) Surgery should therefore be offered to asymptomatic cases at the time of diagnosis. There is little doubt that without surgery, in the following years, symptoms would appear and surgery then would impose additional difficulties as it would then be a much bigger operation.
with a task to remove a bigger piece of lipoma before the cord is well freed. Recovery from an advanced deficit is always more difficult and incomplete. After thorough neuroimaging and urological work up, surgery can be done between 3 to 6 months old. In a retrospective study done in Tuen Mun Hospital, better postoperative results are obtained for those done early as opposed to those diagnosed late and surgery done after 4 year old.\textsuperscript{10}

The prime target is to free the whole spinal cord from any tethering within the dura. (Fig. 6,7) This usually means that the lipoma, which spans between the dorsal root entry zones on the sides, has to be completely excised along a silvery plain between the lipoma and the neuroplacode. Cutting along the precise plain with scissors is a far better way than using the surgical aspirator or lasers. At the end of excision, in transitional lipoma in particular, as the placode is flattened by the adherent lipoma, it has to be neurulised - the placode stitched together forming a cord like structure again. This is an effective way to avoid adhesion and thus retethering.

Proper meticulous watertight closure follows. Unlike open lesions, skin closure is seldom a problem. It is not necessary to pay special effort to excise extradural aberrant tissue except skin sinus and appendages. Cutaneous haemangiomas usually vanish with time. Similar nursing precaution is to be taken after surgery as in open cases.

For the rare incidence of split cord malformation (SCM), the principle is similar in that the midline septum in between the 2 hemicords has to be excised totally. For Type I SCM in which each hemicord has its own dural sheath, the 2 dural sheaths have to be opened and stitched as one alleviating the possibility of dural tethering later on.

**Complication**

Wound healing problems leading to CSF leakage and meningitis remain the most feared complication. However with appropriate precautions, such occurrence can be avoided totally. With effective multiple layer closure, subcutaneous pseudomeningocele is noted in about 5% of cases. With time, most of them would subside spontaneously. Additional deficit is rare after surgery.\textsuperscript{10}

**Results**

For symptomatic cases, pain is the most ready to improve irrespective of the pathology - be it just a fatty tight filum or a transitional lipoma.

Sensorimotor deficit comes next in terms of responsiveness. Acute recent deficit recovers better than those old burnt out deficits with orthopaedic deformities.

Bladder dysfunction also benefit apart from those atonic bladders with large residual volume which probably would require intermittent catheterisation indefinitely.

The general principle holds here in that the more advanced the disease is on presentation, the worse they would fair despite the best treatment.

**Conclusion**

Spinal dysraphism is one of the most common causes of disability in infants and children. It can be substantially reduced with folate and better dietary habits. Together with better antenatal care, the incidence is on the decline for the open type. However, occult lesions still require concerted efforts of clinicians in staying vigilant to make the right diagnosis, treatment instituted promptly with surgical expertise and followed with multi-disciplinary approach to get the best result.
References


MCHK CME Programme Self-assessment Questions

Please read the article entitled “Spinal Dysraphism” by Dr. Dawson Fong and complete the following self-assessment questions. Participants in the MCHK CME Programme will be awarded 1 CME credit under the Programme for returning completed answer sheet via fax (2865 0345) or by mail to the Federation Secretariat on or before 31 December 2006. Answers to questions will be provided in the next issue of The Hong Kong Medical Diary.

Questions 1-10: Please answer T (true) or F (false)

1. With the advent of sophisticated neuroimaging modalities, the incidence of open spinal dysraphism in neonates is on an increase.

2. Folate as a vitamin supplement during pregnancy is not good enough to prevent dysraphism in the embryo.

3. The more rostral a dysraphic defect is, the more extensive is expected of the morbidity.

4. Children with cutaneous stigmata along the midline need to be investigated with spinal MR irrespective of whether there is demonstrable neurological deficit.
5. Severity and extent of occult dysraphic defect is directly proportional to the neurological deficit at birth.

6. Hydrocephalus is commonly associated with occult dysraphism.

7. For a baby born with myelomeningocele, the defect should be well dressed to prevent CSF leak.

8. Surgery can be delayed until the baby is older and stronger.

9. As the patient with an occult lesion grows and gets taller, new symptoms would arise from repeated mechanical damage as well as a worsening tethering on the spinal cord.

10. There is no difference when to operate on an occult dysraphism in terms of technical difficulty and patient outcome.

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**Answers to November 2006 issue**

Early Diagnosis of Spondyloarthropathies

1. b  2. c  3. a  4. b  5. d  6. a  7. c  8. c  9. d  10. b

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