SPLIT NOTOCHORD SYNDROME ASSOCIATION

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CASE PRESENTATION :-

New born baby, boy, referred to the paediatric surgical team at the age of 14 hours.

Birth History :-

Born in Al Wasl Hospital at term by NVD with B.W. of 3.760 kg. Apgar score was 8 & 9 at 1 & 5 min respectively.
Maternal History :-

Mother is 29 years old. P2+1.

Antenatal USS was normal. No history of polyhydramnios.

The first baby died at the age of 6 months because of cardiac abnormality.
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ON EXAMINATION :-

After birth, the baby became tachypneic, grunting, cyanosed for which he was transferred to the SBCU, intubated and connected to a mechanical ventilator.

The initial blood gases showed pH:7.324, PO$_2$:40.2, PCO$_2$:46.7, HCO$_3$:23.6.
GENERAL EXAMINATION :-

The baby was on ventilator with high parameters, pink in color, no pallor, no dysmorphic features, no skin lesions.

Head & neck examination was normal.

Chest examination revealed good bilateral air entry but it was more on the left side.

Cardiovascular system was normal.
Abdomen was soft, scaphoid, no palpable masses, no organomegaly.

Locomotor system was normal.

External genitalia – Male with bilateral impalpable testicles.

Anus & perineum was normal.

Chest x-ray revealed bowel gas shadow at the right hemithorax and Bifid Dorsal Spine.
Bowel shadows in the left hemithorax.

Bifid upper thoracic spine.
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INVESTIGATIONS :-

Initial blood tests was normal and new blood gases showed pH:7.5, PO$_2$:36, PCO$_2$:30.3, HCO$_3$:24.9.

Upper GI contrast study showed the contrast passing through the small bowel and filling the right hemithorax with diagnosis of right congenital diaphragmatic hernia.
Upper GI contrast Study showing Bowel loops in The left hemithorax
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Treatment :-

The baby underwent surgery on the same day through right sub-costal incision.

The liver was mobilized and a mid-ileal loop was seen herniating through a defect in the root of the mesentery.

It was difficult to reduce all the bowel loops.
Small bowel was resected and primary end to end ileal anastomosis was done.

The herniating intestine was delivered in pieces and the defect was closed.

No defect in the right diaphragm was seen.

The wound closed in layers.
Post operative x-ray
Showing all bowel loops Reduced to the abdominal Cavity. Normal right dome Of the diaphragm
Post operative period :-

Uneventful apart from one attack of convulsion on the 9th post operative day.

Then the baby discharged home in good general condition on 19th Jan ’02.

He attended clinic appointment after one week and was doing well.
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The first x-ray impression was right diaphragmatic hernia with completely split upper thoracic spine and this abnormality could be described as thoracic spine split notochord with anterior thoracic meningocele or diastematomelia for which CT or MRI of the thoracic spine was suggested.
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On (MRI) → Spinal dysraphism in the form of:

- Upper dorsal scoliosis,
- Spina bifida occulta of C7 down to T6,
- Butterfly T3, T4 and T5 vertebrae.
- Diastematomelia of the upper dorsal cord with
- No MRI evidence of bony or connective tissue,
- Finally, anterior thoracic meningomyelocele.
DIASTEMATOMYELIA (DM)

DM is a rare form of spinal dysraphism characterized by a sagittal cleft of varying extent in the spinal cord, conus medullaris and filum terminale with splaying of the posterior vertebral elements.

This condition is the result of the presence of an osseous, cartilaginous or fibrous septum producing a complete or incomplete sagittal division of the spinal cord into two hemicords.
In about 50% of patients, each hemicord contained within its own dural tube and separated by a dura-sheathed rigid median septum, while in the remaining part they lie within a single dural tube separated by a non-rigid, fibrous median septum as seen in our patient.

Associated intra-medullary tumours with a DM have been rarely described.
DIASTEMATOMYELIA (DM)

- It may be isolated or associated with other segmental anomalies of the vertebral bodies.
- DM usually occurs between T9 and S1 levels.
- Cervical DM is a very rare entity.
- There is a female predominance in these patients and it was more remarkable in Type I DM than in Type II DM.
DIASTEMATOMYELIA (DM)

Pang and his colleagues have suggested an alternative classification to deal with all double spinal cord malformations as a split spinal cord malformations with Type I and Type II split spinal cord malformation (SSCMs).
Initially, the embryo has two layers – endoderm & ectoderm.

Mesoderm forms between the two, but for a short while, at the primitive pit, the two layers remain in contact.

A transient opening, the neureneretic canal, appears connecting the neural ectoderm with the gastro-intestinal endoderm.
DIASTEMATOMYELIA (DM)

The notochord forms in the mesoderm just caudal to the neurenteric canal.

As it migrates, the notochord is split by the persistent neurenteric canal, resulting in the development of spina bifida or other vertebral anomalies and anterior or posterior myelomeningocele.

The term *split notochord syndrome* has been applied to the defect.
POSSIBLE ANOMALIES RELATED TO FAILURE OF REGRESSION OF THE NEURENTERIC CANAL ARE :-

- Complete dorsal enteric fistula
- Fibrous cord passing through the spinal cord as in DM.
- Intra-spinal enteric cyst.
- Dorsal enteric sinus
- Neurenteric cyst and
- Enteric duplications.
SYMPTOMS

- Skin stigmata
- Orthopedic deformities of foot.
- Spina bifida aperta
- Weakness in lower extremities
- Scoliosis
- Bladder and bowel disturbance
- Short &/thin leg
- Back pain
**DIASTEMATOMYELIA (DM)**

<table>
<thead>
<tr>
<th>Skin findings</th>
<th>Orthopedic deformities</th>
<th>Neurological findings</th>
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<tr>
<td>Hypertrichosis</td>
<td>Scoliosis</td>
<td>Paraparesis</td>
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<tr>
<td>Capillary haemangioma</td>
<td>Kyphosis</td>
<td>Unilateral leg paresis</td>
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<tr>
<td>Hyperpigmentation</td>
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<tr>
<td>Subcutaneous lipoma</td>
<td>Pes cavus/valgus</td>
<td>Bladder and bowel dysfunction.</td>
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<tr>
<td>Dimple</td>
<td>Trophic ulcers</td>
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RADIOLOGICAL FINDINGS

Plain x-ray

- Bifid lamina.
- Scoliosis.
- Hemi vertebra.
- Accessory lamina.
- Fused ribs.
- Widened interpediculate distance

- Bony median septum.
- Bifid vertebra.
- Kypho-scoliosis.
- Sacral agenesis.
- Blocked vertebra.
**MRI FINDINGS**

♫ Splitting of the spinal cord and other dysraphic lesions such as myelomeningocele, meningocele and lipoma.

♫ Low conus medullaris, thick filum terminale, hydromelia, split vertebrae and rigid median septum.
DIASTEMATOMYELIA (DM)

CT Myelography was superior to other radiological tools in defining the type of DM.

Prenatal and post natal ultrasound can be of diagnostic value.
ASSOCIATED LESIONS

85% of the patients had more than one spinal lesion as

- Thick filum terminale.
- Myelomeningocele.
- Meningocele.
- lipo-myelomeningocele.
- intra-dural arachnoid cyst.
- dermal sinus tract.
- dermoid cyst.
- teratoma.
- dorsal lipoma.
DIASTEMATOMYELIA (DM)

Treatment :-

When diagnosed, all cases of DM should be surgically treated even if the patient was neurologically asymptomatic.
A good outcome is expected aiming at improvement or stabilization of the deficit.

COMPLICATIONS

- Transient.
- Permanent.
Pathological examination of the specimen from the median septa revealed:

* Fetal renal tissue
* Lymphoid tissue
* Dermoid cyst
* Muscle tissue
* Ganglion and blood vessels.
* Bone and cartilage.
DIASTEMATOMYELIA (DM)

Endodermal, ectodermal and mesanchymal structure detected within the median septa support the theory of endomesanchymal tract and ectoendodermal adhesion.
THANK YOU