IDIOPATHIC SYRINGOMYELIA-
ALDER HEY EXPERIENCE

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CONFLICT OF INTEREST DISCLOSURE

• Grants/research support- none
• Consultant- none
• Stock/shareholder- none
• Royalties- none
• Other financial support- none
• Employee- none
IDIOPATHIC SYRINGOMYELIA

• No identifiable underlying cause like Chiari, tumor, trauma, etc.

• Underlying pathologies discovered till date-
  - difficult labour*
  - arachnoid webs, pouches, cyst**
  - small posterior fossa [Chiari O]***
  - abnormal CSF flow dynamics****

**** Struck AF, Haughton VM. Idiopathic syringomyelia: phase Contrast MR CSF flow dynamics at level of foramen magnum. Radiology, October 2009; 253:184-190
IDIOPATHIC SYRINGOMYELIA

Even with increasing number of cases of idiopathic syringomyelia being reported, knowledge about its natural course remains unclear & there is a lack of common consensus regarding treatment and follow-up requirements.

OUR STUDY

- Retrospective and prospective analysis
- 30 cases of Idiopathic syringomyelia from our database (2000-2011).
- Largest study group to date.
- Assessment of - records,
  - clinical symptoms,
  - response to any treatment given and
  - serial MRI imaging at regular intervals (apprx 1yr) has been performed for each patient.
  - Avg. follow up - 4yrs
• **Measurements- on MRI**

1. **Length x width of syrinx**
   - T2W sag

2. **Syrinx length/vertebral body height ratio (SL/VB)**
   - T2W sag

3. **Max syrinx dia/ Spinal cord dia ratio (SD/SpD)**
   - T2W axial
OBSERVATIONS

SEX DISTRIBUTION
- Male: 17 (57%)
- Female: 13 (43%)

AGE DISTRIBUTION 3yrs-18yrs
- 0-5 yrs: 2 (7%)
- 6-10 yrs: 10 (33%)
- 11-15 yrs: 9 (30%)
- 16-20 yrs: 16 (51%)

LOCATION
- Thoracic: 66%
- Cervical: 22%
- C5-Conus: 6%
- Conus: 6%
OBSERVATIONS

- BACKACHE
- URINARY SYMPTOMS
- INCIDENTAL FINDING
- LEG PAINS
- CAVO VARUS
- GROSS MOTOR DELAY

HYPOTONIA
HEMIATROPHY LEFT LL
HYPOSTHESIA
BRISK KNEE JERKS
NONE

SPINEWEEK 2012  RAI AMSTERDAM  28 MAY - 1 JUNE
<table>
<thead>
<tr>
<th>Location</th>
<th>P. complaints</th>
<th>Clinical signs</th>
<th>Size</th>
<th>MRI</th>
<th>Surgery</th>
<th>Outcome</th>
<th>Post Op size</th>
<th>Sy dia/Sp dia ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>C6-C7</td>
<td>Backache, leg pain, frequency</td>
<td>None</td>
<td>32 x 2</td>
<td>Normal position Conus M</td>
<td>FT Division</td>
<td>Clinical improvement</td>
<td>0.2</td>
<td>0</td>
</tr>
<tr>
<td>Conus</td>
<td>Urgency, frequency, backache</td>
<td>None</td>
<td>27 x 1.5</td>
<td>Normal position Conus M</td>
<td>FT division</td>
<td>Clinical improvement</td>
<td>27 x 1.5</td>
<td>0.2 0.15</td>
</tr>
<tr>
<td>T10-L1</td>
<td>Incidental MRI finding</td>
<td>None</td>
<td>26.5 x 3-42 x 5</td>
<td>Normal position Conus M</td>
<td>FT Division</td>
<td>Under follow up</td>
<td>0.4-0.6</td>
<td>Post-op MRI awaited</td>
</tr>
<tr>
<td>C4-T8</td>
<td>Backache, Urgency</td>
<td>Hyposthesia L/L</td>
<td>164 x 12</td>
<td>Crowding of PF</td>
<td>FMD</td>
<td>Clinical improvement</td>
<td>154 x 5</td>
<td>0.7 0.58</td>
</tr>
</tbody>
</table>
RESULTS

1. No change in SD/SpD ratio - 18
   - Symptomatic improvement - 14 (77.7%)
   - No change - 4 (22%)

2. Increased SD/SpD ratio - 2
   - Deterioration - 1 (50%)
   - No change - 1 (50%)

3. Decreased SD/SpD - 10
   - Symptomatic improvement - 9 (90%)
   - No change - 1 (10%)

DECREASED SD/SpD RATIO GROUP -
- Avg. Follow up - 5 yrs
- Avg. decr. in ratio - 0.1
- 90% (9/10) showed clinical improvement
- 3 patients received surgical treatment -
  - FMD - 1
  - FT division - 2
- 60% (6/10) showed no change in syrinx size
CONCLUSION

• We believe that group of truly idiopathic syringomyelia is shrinking.
• “Treat the patient, not the scan” - Consider division of F T if symptoms fit even if scans may not.
• Early, pre-pubertal FT division/FMD should be considered esp. in patients with scoliosis
• Size of syrinx may not be the only radiological parameter to guide treatment and follow-up.
• Follow up may only be required until SD / SpD ratio decreases or symptoms improve.
• There is a need for further Large multi-institutional prospective studies.