ORTHOPAEDIC CONDITIONS IN MOEBIUS SYNDROME

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OUTLINE

- Review of recent Orthopedic literature
- Clubfoot
- Limb Deficiencies
- Scoliosis
POSSIBLE ORTHOPEDIC FINDINGS
Orthopedic Manifestations of Mobius Syndrome: Case Series and Survey Study

Philip McClure,¹ David Booy,¹ Julia Katarincic,² and Craig Eberson³

Survey of Foundation Members 2013-2014
96 respondents

Table 3: Patient survey results.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Number</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hand:</td>
<td></td>
<td></td>
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<tr>
<td>Difference in digital anatomy</td>
<td>35</td>
<td>36.5%</td>
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<tr>
<td>Syndactyly</td>
<td>17</td>
<td>17.7%</td>
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<tr>
<td>Upper extremity nerve deficit</td>
<td>28</td>
<td>29.2%</td>
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<tr>
<td>Foot:</td>
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<td></td>
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<tr>
<td>Difference in digital anatomy</td>
<td>17</td>
<td>17.7%</td>
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<tr>
<td>Syndactyly</td>
<td>6</td>
<td>6.3%</td>
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<tr>
<td>Clubfoot</td>
<td>40</td>
<td>41.7%</td>
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<tr>
<td>Congenital vertical talus</td>
<td>3</td>
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<tr>
<td>Planovalgus foot</td>
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<td>38.5%</td>
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<tr>
<td>Chest:</td>
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<td></td>
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<tr>
<td>Missing/weak pectoralis</td>
<td>29</td>
<td>30.2%</td>
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<tr>
<td>Missing/weak back musculature</td>
<td>14</td>
<td>14.6%</td>
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<tr>
<td>Sternal abnormality</td>
<td>23</td>
<td>24.0%</td>
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<tr>
<td>Spine:</td>
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<td></td>
</tr>
<tr>
<td>Scoliosis</td>
<td>27</td>
<td>28.1%</td>
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<tr>
<td>Kyphosis</td>
<td>10</td>
<td>10.4%</td>
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<tr>
<td>Lordosis</td>
<td>5</td>
<td>5.2%</td>
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<td>Missing bone in spine</td>
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<td>2.1%</td>
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<tr>
<td>Difficulty with anesthesia</td>
<td>26</td>
<td>27.1%</td>
</tr>
<tr>
<td>Positive family history</td>
<td>5</td>
<td>5.2%</td>
</tr>
</tbody>
</table>
Mobius Syndrome: A 35-Year Single Institution Experience

Philip K. McClure, MD, Eray Kilinc, MD, Scott Oishi, MD, Anthony I. Riccio, MD, and Lori A. Karol, MD

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- Texas Scottish Rite Hospital, Dallas TX
- 44 patients (over past 35 years)
- Compared to general population, increased incidence of:
  - Clubfoot (41%)
    - 13/18, 72% bilateral
    - 12/18, 67% required surgery*
  - Hand deformity (25%)
  - Poland Syndrome / Chest wall abnormalities (20%)
  - Scoliosis (14%)
Clubfoot

Congenital talipes equino-varus
- The most common & significant congenital deformity worldwide

Incidence by race:
- Caucasians: 1 per 1000 live births
- Africans: 2-3 per 1000
- Polynesians: 6 per 1000
Clubfoot Facts

- Male : female ratio 3:1
- 40% bilateral
- 2nd child in family with clubfoot = 1/35
- Monozygotic twins 32.5% coincidence
- Dizygotic twins 2.9%
- Probably multifactorial condition with polygenic antecedents
- Associated with smaller calf musculature / atrophy on affected side likely due to decreased innervation
CLUBFOOT ETIOLOGY

- Developmental arrest / delay
  - Fetal foot passes through equino-varus stage

- Intra-uterine molding
  - Molding can cause equinovarus position, but this is easily correctable and occurs late in pregnancy

- Retracting fibrosis / myofibroblast activity
  - Collagen / myofibroblasts, similar to Dupuytren’s
  - Cause / effect?

- Neuromuscular
  - Neuromuscular tissues of the calf
  - Association also seen with spina bifida / CP
Cavus (midfoot): Increase of the height of the medial arch of the foot

Adductus (forefoot) / Varus (hindfoot)
- distal part moves towards median body plane

Equinus (hindfoot): Increased plantarflexion of the foot
Severe tibio-talar plantar flexion
Medial talar neck inclination
Severe medial displacement of navicular
Wedge shaped navicular
Wedge shaped head of talus
Adducted and inverted calcaneus
Wedge shaped distal calcaneal articular surface.
Medially displaced cuboid
TREATMENT: THE PONSETI CASTING METHOD

Clubfoot treatment over 4 – 6 weeks

Stage 1  Stage 2  Stage 3  Stage 4  Stage 5
Bracing Afterwards is ALWAYS Required

Clubfoot loves to recur

- Ponseti braces or Dennis-Browne bar
- Worn until some point between 3 & 5 years of age
SURGICAL TREATMENT

No longer the first line treatment method!

- Only indicated when casting fails
  - More often in Moebius, arthrogryposis, neurologic deficits, etc.

- 50% of surgical feet need a 2\textsuperscript{nd} procedure

- Occasionally after casting a child may have some residual “dynamic supination”
  - May be indicated for Anterior Tibialis Tendon transfer at age 3 or 4.
**SURGICAL OPTIONS**

**Posteriomedial-Lateral Release**
- Leads to stiffness
- May need to be redone

**Anterior tibialis tendon transfer**
- Turns inverter to everter
Salvage Surgery Options

Taleectomy
- Removes bone that blocks correction
- Stiffness
- Best for patients who don’t walk a ton

Triple Arthrodesis
- Removes portions of bone and allows fusion in new position
- Used in neglected / resistant clubfeet closer to maturity
HAND ABNORMALITIES

- Missing, shortened or fused fingers (syndactyly)
- Most often do not require treatment other than possible syndactyly release
Poland Syndrome

- Unilateral absence of pectoralis muscle
Poland Syndrome

- Physical Therapy
- Cosmetic Surgery
TRANVERSE LIMB DEFICIENCY
Scoliosis

- 3 Dimensional curvature / rotation of the spine
- Idiopathic: Unknown cause
- Neuromuscular

- Often appears close to adolescence
**Definition of Scoliosis**

- Curve measuring over 10 degrees on X-ray
- Curves over 25 degrees respond to bracing
- Curves over 45 degrees typically require surgery to prevent progression and arthritis in adulthood
Scoliosis Surgery
SUMMARY

- Spectrum of Orthopedic Issues associated with Moebius syndrome
- Clubfoot is most common limb condition (40%)
  - Neuromuscular etiology
  - Always try Ponseti casting first!
- Finger, Limb, Chest wall deformities
- Scoliosis
  - Treat as idiopathic scoliosis: brace if early (25-45 deg)