Congenital Anomaly of The Upper Extremity
(2004, ICL, KPOS)

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Sprengel’s Disease
(Congenital high scapula)
Sprengel’s disease

Pathology

- **Scapula**
  - high, small (smaller vertical diameter and apparently greater width)
  - Inf. angle: medial rotation
  - Sup. angle: tilted forward.

- **Clavicle**
  - tilted toward upward and laterally
Sprengel’s disease

Pathology

- **Omovertebra**: connection between the spinous process and scapula bony, cartilaginous, fibrous

- **Musculature**:
  - *Trapezius*: most often affected
  - rhomboid, levator, serratus ant: hypoplastic and fibrotic
Sprengel’s disease

Clinical Feature

- F : M = 3 : 1
- Left is more often
- 3-5 cm higher in average
- Abduction is limited
  GH joint : Normal
  TS joint : restricted
Treatment

considering factors

1. severity of deformity
2. functional impairment
3. associated anomaly
4. age
# Sprengel’s disease

## Severity of deformity

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>very mild: invisible in cloth</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>mild: lump in undressed</td>
<td>simple excision</td>
</tr>
<tr>
<td>3</td>
<td>moderate: elevated 2-5 cm</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>superior angle near the occiput</td>
<td></td>
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</table>

3, 4: scapula should be lowered
Sprengel’s disease

Functional impairment

- Omovertebra bone: resection of the omovertebra
- Motor weakness caused by fibrosis, hypoplasia
Sprengel’s disease

**Associated anomaly**

- Klippel-Feil syndrome
- Kyphosis
- Scoliosis

: overshadow the deformity
Sprengel’s disease

Age (Op timing)

3-7 years

• < 3 : extensive and difficult
• > 7 : greater danger of brachial plexus injury
• recently, 6-9 months ? (Tachdjian)
Sprengel’s disease

Technique

Numerous!
: Most popular

• Woodward
  • Green
Treatment

extensive surgery

• normal anatomic appearance cannot be provided!
• caution must be exercised!
• expectations should be realistic!
Sprengel’s disease

Treatment

Green Technique

• divided at periscapular area (scapular insertion)
• traction wire (scapula to ilium)
• Pro: definite improvement of function
• Con: difficult procedure
Sprengel’s disease

Treatment

Woodward Technique

- detaching the origin of the trapezius and rhomboid muscle from spinous process
- Pro: more easier (but not not easy)
- Best result
high scapula due to neurogenic spasm
Case of Grade 2
Case of Grade 4
Cho et al found ---3DCT is can be helpful in delineating the deformity and planning scapuloplasty.

10th edition, Campbell’s Operative Orthopedics, 2003
Retracted omovertebra bone
Post. Aspect of Transverse process
Spinous process
Preop x-ray

Sprengel’s Disease
2, Female,(1540231)

Preop clinical
Op (Woodward) at 4 Y, Postop X-ray

Follow up clinical photo

Female,(1540231)
Congenital Muscular Torticollis (CMT)
Etiology

exact etiology is not known
1) compartment syndrome
2) in utero crowding
3) neurogenic
4) mesenchymal cell
Clinical Finding

Head tilting : involved side
Chin rotating : the opposite side
mass : maximum within the first 4 weeks, then gradually regressed

7-20% combined with DDH or acetabular dysplasia
Torticollis

Incidence of Combined DDH

20% ?

8% : Walsh JJ
(JPO,1998, 219-21)

11.6% : Cheng JC
(J. Pediatr,1999, 134, 712-6)
Torticollis

Treatment

- Initial treatment: conservative
  (effective ?, spontaneous resolution ?)
- Surgical treatment:
  persistent deformity after 1 years
DDX

• Congenital anomaly (osseous type)
  • Basilar impression
  • Atlantooccipital anomalies
  • Unilateral absence of C1
  • Familial cervical dysplasia
  • Atlantoaxial rotatory displacement
Torticollis

DDX

- Trauma
- Inflammatory
- Ophthalmic problem
Torticollis

**DDX**

- Neurogenic
  - Cervical cord tumor
  - Arnold Chiari syndrome
  - Ocular pathology
  - Proxysmal torticollis of infancy
- Sandifer syndrome
Torticollis due to?
Torticollis due to Kilppel-Feil syndrome
Torticollis due to Spine deformity (Klippel-Feil syndrome)
Atlantoaxial rotatory displacement
Torticollis due to Ophthalmic problem
Torticollis

Treatment

1. Unipolar: mild & moderate
2. Bipolar: older age, recurred, severe case
3. Middle 1/3 resection
4. Complete resection: rare
Torticollis

Treatment

avoid damage of spinal accessory nerve, jugular vein, carotid vessel, facial nerve
Torticollis

Treatment

(post operative care)

cervical collar?
plaster cast?
Buckminster Brown brace?
Torticollis orthosis?
Congenital Muscular Torticollis

preop  postop  Follow up

6, female
(1794787)
unifocal myotomy in old age
unpolar myotomy in old age
unipolar myotomy in old age
unipolar myotomy in old age
1, male
(1707176)
Cong.R-U Synostosis

Clinical feature

• mainly proximal end
• discovers early adolescence
• All most all fixed in pronation: limited supination and pronation
• bilaterality: 60%
Clinical feature

- **Type I**: complete synostosis
  radius is longer and arches anteriorly

- **Type II**: less involved with dislocation of the head
  often unilateral & other anomalies
  (absence of thumb, poly or syndactyly)
Pathology

- initially osteochondrosis, later the osseous bridge
- Narrow & tight interosseous memb
- Abscence of brachioradialis, pronator teres, pronator quadratus, supinator
usually **not necessary** if patient has mild unilateral deformity or no major functional loss
**Procedure to restore motion**

1. Resection of syostotic radius
2. Division of interosseous membrane
3. Synostotic resection + interposition of fat, fascia, silicone
4. Separation + vascularized fat-fascio graft + index procedure (osteotomy of radius)

“Not justified in text book until now”
Treatment

The only useful surgery

osteotomy though the site of the synostosis
Indications for surgery

- Bilateral involvement
- Severe pronation (more than 60)
<table>
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<tr>
<th>Position</th>
<th>dominant</th>
<th>nondominant</th>
</tr>
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<tbody>
<tr>
<td>10-15 pro</td>
<td>Neutral</td>
<td>45 sup</td>
</tr>
<tr>
<td>30-45 pro</td>
<td>Neutral</td>
<td>45 sup</td>
</tr>
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</table>
Complication

Compartment syndrome

- 1/3
- common in osteotomy with greater than 85 degree of correction
Congenital Radio-ulnar Synostosis

6Y, male
(1927280)
6Y, male
(1927280)
LOM on pronation & supination
Congenital Radial Head Dislocation (CRHD)
Clinical feature

• Most common congenital anomaly of the elbow
  • Ant:47%, Post:43%, Lateral:10%
  • do not noted until 4-5 years of age (or school age)
DDX

acquired traumatic form
(missed Monteggia, Fx of radial neck, pulled elbow)
DDX

1. bilaterality, familial Hx, trauma Hx, associated anomaly
2. Radiologic findings
   capitulum: small, hypoplastic (primary)
   radial head: ovoid, small (secondary)
   radial notch of ulna: small or absence
Post-traumatic D/L  
Neglected Monteggia

Congenital D/L
Cong.D/L.Radial Head

Inheritance

- Autosomal dominant or X-linked recessive
- Associated multiple exostosis, nail patella syndrome, antecubital pterygium syndrome
  (60% with other syndrome)
Congenital radial head dislocation with camptodactyly
Congenital radial head D/L with clubhand (thumb hypoplasia)
Congenital radial head D/L with clubhand (thumb hypoplasia)
Congenital radial head D/L with clubhand (hypoplasia of thumb)
Congenital radial head dislocation with Pterygium syndrome
Congenital radial head dislocation with Pterygium syndrome
Associated with osteochondroma
Treatment

• Surgical treatment is seldom necessary in childhood

• Radial head resection before skeletal maturity has been associated with several different complications

(By Chapman's Orthopedic Surgery, 2nd edition & Campbell's Operative Orthopedics, 10th edition)
Treatment

younger than 1-2 y

lengthening of the ulna
+ shortening, osteotomy of radius
+ reconstruction of the annular ligament

: preliminary procedure
Treatment

Older than Infant
Resection of the radial head

1. Progressive pain
2. Progressive LOM
3. Progressive Limitation of activity

Regardless of Age!
(common in trauma, but rare in congenital, )
Treatment

open reduction in the infancy controversy!

• no evidence to justify?
• recommend until 3 years?
Anterior dislocation
Congenital D/L of Radial Head

female, both

(1748019)
Reconstruction of annular ligament
Arthrogryposis
Arthrogryposis

Clinical feature

not specific disorder but a symptom complex associated neurogenic and myopathic

1) multiple symmetric joint contracture
2) atrophied extremities
3) marked limitation of motion
Clinical feature

More severe distally than proximally
Most upper extremity is affected
### Clinical feature

**Deformity of upper extremity**

- **shoulder**: adducted and Int. rotated
- **elbow**: fixed in extension: (most limiting deformity)
- **forearm**: pronated
- **wrist**: wrist flexion with ulnar deviation
- **fingers**: gathered together, thumb in palm
Clinical Feature

Gather together
Thumb in palm

Internal rotation & adduction

Fixed in extension
Pronated
Flexion with ulna deviation

Arthrogryposis
(1824618)
Arthrogryposis (1824618)
Arthrogryposis

Ultimate goal of Treatment

• Independent upper extremity function for allow for activity of daily living
  1) bring to the hand to mouth for hygiene and feeding
  2) can be use for pushing up to a sitting position
  3) using an ambulatory aid, such as a crutch
Arthrogryposis

**Treatment**

PT, casting, splinting

Goal: passive elbow flexion by 2 y (6 months) of age

- Obtained
- Not obtained

No active motion until 5 Y (4Y) → Triceps lengthening & capsulotomy

Tendon transfer (Ticeps to Bicep)
Arthrogryposis

Treatment

• Timing of treatment: neonatal period

• ROM exercise and stretching followed prolonged splint

• normal intelligence & sensate skin

• marked adaptability and surgery is often unnecessary
<table>
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<th>Arthrogryposis</th>
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<tr>
<td><strong>Treatment</strong></td>
</tr>
<tr>
<td>surgical release</td>
</tr>
<tr>
<td>(no response to adequate trial of 6 months to 1 year)</td>
</tr>
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**Arthrogryposis**

**Treatment**

**Elbow**

Dynamic or static splint: until 40-50 flex.
Older 2 years: elastic harness
No passive motion: post. capsulotomy & lengthening of triceps
Good hand and wrist function: Steindler flexoplasty
Arthrogryposis

Treatment

shoulder

- active and passive stretching
- **External rotation osteotomy** at proximal 1/3 (if it can not approximate 90 degree ext. rotation)
- shoulder fusion is not necessary
Arthrogryposis

Treatment

Wrist

• volar capsulotomy & tendon transfer
  (FCR & FCU to the dorsum of wrist)
• rigid type: prox. row carpectomy with tendon transfer

Finger

• casting & splinting
• surgery seldom improve the function
Congenital Transverse failure of forearm
Congenital Transverse Failure of Forearm (congenital amputation)
Congenital Transverse Failure of Forearm with Congenital Radial head
Characters

• Subclavian artery supply disruption?
• The most common level is proximal forearm
• Unilateral
• Remakably few functional deficit
**Treatment**

**BE or AE type**

**6 months**
Prosthetic team
passive hand or mitt
when able to sit independently

**2 to 3 years**
Active terminal device (TD)

**More proximal or distal failure**

No prosthesis in Infancy

Body powered (cable-operated)

Myoelectric prosthesis

* Krukenberg forearm: rarely indicated
Congenital Elbow Synostosis
Clinical Features

• Humeroradial > Ulnotrochlea
• No forearm motion
  (in position of 60-90 flexion ?)
• Associated upper limb malformation
  (ulnar clubhand, phocomelia)
Treatment

Dysfunctional position: corrective derotational osteotomy

- Excision & motion restoration procedure
- Continuous passive motion device

Not improved results
Humeroradial synostosis
+ Ulnotrachlear synostosis
+ Ulnar clubhand
+ Hand: agenesis of 5th metacarpal bone
Congenital Clubhand
(congenital radial deficiency)
Clinical feature

- forearm bowed to the radial side with prominent distal radius
- 2/3 of the length of the opposite unaffected extremity
Clinical feature

- **Thumb**: defective or absence
- **Thenar muscle**: defective in proportion to the deficiency of the thumb
- **Flexor**: present but not well differentiated distally
- **Extensor**: deficiency at its origin
- **Neurovascular**: absence of radial A.N
Classification

• **Type 1:** short distal radius: dist. epiphysis is present but suppressed: Tx is confined to the thumb deficiency

• **Type 2:** hypoplastic radius: dist. & prox. epiphysis is present, but growth is defective in both area; Tx would be directed toward the lengthening of the radius, but not found to be effective
Classification

- **Type 3:** partial absence of the radius: supplying support of the hand
- **Type 4:** total absence of the radius: release of the soft tissue and provide support of the hand: **most common**

* **TAR syndrome:** thrombocytopenia + complete absence of radius
Congenital Clubhand, Type 1
Congenital Clubhand, Type 1
Congenital Clubhand, Type 2
Congenital Clubhand Type 2
Congenital Clubhand Type 2
Congenital Clubhand
Female, 1 month, type 4 (1731114)
Associated Anomaly

- rarely occur as a isolate (40 % : unilateral., 77 % : bilateral --associated anomaly)
- **Fanconi syndrome** (radial defect + severe aplastic anemia)
- **TAR syndrome**
- **VATER syndrome** (ventricular or vertebral anomaly + T-E fistula + renal anomalies)

→ **VACTERL/S SYNDROME**
Treatment from no treatment to aggressive correction
**Treatment**

main shortcoming
1) lack of wrist stability
2) impaired finger motion
3) short forearm
4) absence and defective thumb
Treatment

1) bone graft
2) fusion of the wrist
3) Centralization: Tx of choice
4) lengthening of the radius
5) epiphysiodesis
6) Radialization with tendon transfer
Cong. Clubhand

Current Treatment

1. initial treatment
   at birth: serial casting
   **definite surgery : 6 months- 1 years**
   1) proper preoperative splinting, stretching and tendon transfer
   2) osteotomy of the ulna : bowing >30
   3) thumb reconstruction
2. 1-3 years old who first present: **external fixator** to stretch tight radial structure (casting will fail in this age group)
Congenital ulnar clubhand (Ulnar dysplasia)
Type 1: Hypoplasia of the ulna + hand malformation
Type 2: partial loss of ulna + stable elbow, radius bowing
Ulna

Radial nerve

Elongated, loose capsule