CLEFT HAND & WINDBLOWN HAND
Swanson Classification

- **Typical Cleft Hand**
  - Congenital, Developmental Arrest
    - Longitudinal (vs. Transverse)
    - Central Deficiency (vs. Preaxial, Postaxial, Phocomelia)

- **Windblown Hand**
  - Congenital, Failure of Differentiation
    - Contracture
    - Arthrogryposis
SWANSON CLASSIFICATION OF CONGENITAL UPPER LIMB ABNORMALITIES

I. Failure of Formation of Parts
   Transverse
   Longitudinal
      Radial club hand
      Cleft hand
      Ulnar club hand
      Phocomelia
II. Failure of Differentiation or Separation of Parts
   Synostosis
   Radial head dislocation
   Symphalangism
   Syndactyly
   Contracture
      Arthrogryposis
      Trigger finger
      Clasped thumb
      Camptodactyly
      Clinodactyly
      Windblown hand
      Kirner deformity
III. Duplication
   Polydactyly
IV. Overgrowth
   Macrodactyly
V. Undergrowth
   Thumb hypoplasia
   Madelung deformity
VI. Congenital Constriction Ring Syndrome
VII. Generalized Skeletal Abnormalities and Syndromes
Cleft Hand

- A Longitudinal Deficiency of the Central Rays Results in Cleft Hand
- Cleft Hands are Classified as Typical or Atypical
Atypical Cleft Hand

- Atypical
  - U-shaped defect secondary to absence of the index, long, and ring fingers
  - Unilateral
  - A Form of Symbrachydactyly
  - A Transverse Deficiency of Formation
Typical Cleft Hand

- Typical
  - Contains a Deep V-shaped Central Defect Secondary to Hypoplasia of the Long Ray
  - Usually Bilateral
  - Usually Inherited
Genetic Abnormalities Found

- Mutation of the Split Hand Split Foot gene
  - Localized on 7q21
- Mutation of HoxD 13 gene on chromosome 2q
Inheritance

- Autosomal Dominant Trait, Most Common
- Autosomal Recessive Variant, Less Common has also been described
- 30% of patients with the AD variant are carriers with No abnormalities
- Penetrance is extremely variable amongst affected family members
Clinical Presentation

- Condition may be unilateral or bilateral with or without lower extremity involvement
- Severity ranges from a minor cutaneous cleft to near complete digital loss with preservation of only the small finger
- Syndactyly of the bordering digits is common
- First web contracture is also common
- Transverse phalanges or metacarpals may also be present
Manske Classification

- **Type I:** Normal 1\textsuperscript{st} web
- **Type IIa:**
  - Thumb web mildly narrowed
- **Type IIb:**
  - Thumb web space is severely narrowed
- **Type III:**
  - Thumb and index syndactylized (web space obliterated)
- **Type IV:**
  - Index ray suppressed and thumb web merged with cleft
- **Type V:**
  - Thumb elements suppressed, ulnar rays remain, thumb web no longer present
Surgical Options and Treatment

- Comestic and Functional Outcomes
- Consideration in Treatments Include
  - Deformity (Syndactyly and Transverse Bones)
  - Cleft Deformity
  - 1st Web Space
  - Management of Absent Thumb
  - Other Extremity Deformity (Feet)
- Syndactyly release
  - Redundant cleft skin can be used as skin graft
- Removal of transverse bones
  - Transverse bones keeps the cleft opening
  - Transverse bones often stretch from one MC head to another
  - Care must be taken to preserve or reconstruct damaged capsular and ligaments
- 1st Web Space Release
- Hypoplastic / Absent Thumb
  - The small finger may be pollicized to allow for pinch against a fused post
Windblown Hand
Windblown Hand

- Rare Congenital Ulnar Drift of Fingers with flexion contraction
- First Described by Emile Boix in 1897 as “windblown fingers”
Zancolli & Zancolli described the condition as a type of Segmental Arthrogryposis in 1985
- Developed a Classification System

Wood and Biondi in 1990 coined the entity as “windblown hand”
Arthrogryposis

- Arthrogryposis, or arthrogryposis multiplex congenita, comprises a variety of conditions characterized by multiple joint contractures found throughout the body at birth.
- The condition occurs in 1/3000 live births.
- Prognosis depends on the underlying cause, but most have a normal lifespan.
- If there is a CNS problem, 50% of patients will die in the first year.
- Cranial and CNS, Extremity Displasia and Joint Contractures, Vertebra Displasia
Freeman-Sheldon Syndrome 1938
- Craniocarpotarsal Dysplasia
- Whistling Face Syndrome – Burian 1963
- Craniofacial, Hand, and Foot Anomalies
Clinical Presentation

- Ulnar Drift of the Digits is often present at birth and become more pronounced as the child grows.
- Flexion Contracture of the MP Joint with Ulnar Deviation of the Fingers.
- The Thumb is webbed to the palm by a Soft Tissue Bridge.
- There may also be flexion contractures of the IP joints.
- Deformity is usually Bilateral and Symmetrical.
Conservative Treatments

- Nonsurgical Management in the Form of Splinting
- Requires a great deal of compliance from the child and the parents
- Thumb adduction contracture had been shown to be difficult to correct with splinting alone
- Loss of valuable time in the child’s formative years and interfere with schooling
- Long Time and Suboptimal Result are seen
Surgical Treatments

- Requires Experience and Judgement
- Focus on Contracture Release and Return of Hand Function
  - Addresses Finger Contracture
  - Thumb and First Web Contracture
  - Late Dx and Findings with additional Deformity
    - Relocation of the Subluxed Extensor Tendons
    - Metacarpal Corrective Osteotomy
    - Crossed Intrinsic Transfers
Surgical Management of Windblown Hand


23 Hands in 18 Patients (Age 6 months to 16 years)

Recommends Early Surgical Management for Better Results
<table>
<thead>
<tr>
<th>Type</th>
<th>Deformity</th>
<th>Our series</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Fasciocutaneous deformity, with short skin and abnormal subcutaneous bands</td>
<td>11 hands</td>
</tr>
<tr>
<td>II</td>
<td>Type I + short tendons of the wrist, thumb, and fingers</td>
<td>8 hands</td>
</tr>
<tr>
<td>III</td>
<td>Type I and II + short ligaments and capsules with bony deformity</td>
<td>4 hands</td>
</tr>
<tr>
<td>Procedure</td>
<td>Number of hands</td>
<td></td>
</tr>
<tr>
<td>----------------------------------------------------------------</td>
<td>-----------------</td>
<td></td>
</tr>
<tr>
<td><strong>Fingers</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Contracture release and skin graft</td>
<td>19</td>
<td></td>
</tr>
<tr>
<td>Extensor tendon relocation</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>Metacarpal corrective osteotomy</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Crossed intrinsic transfers</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td><strong>Thumb</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Release and dorsal rotation flap</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>Release and index transposition flap</td>
<td>13</td>
<td></td>
</tr>
<tr>
<td>Metacarpophalangeal arthrodesis</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Skin graft</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>
Flexion Contracture of Fingers

- Transverse Relaxing Incision Just Distal to MP joint Volar
- Subcutaneous Tight Bands were released
- Full Thickness Skin Graft
- Position was Maintain with K wires
- Centralization of Extensor Tendons was achieved by plication of the radial sagittal bands
- Ulnar drift was corrected by means of crossed intrinsic transfer
- Ulnar intrinsics were released at the level of the MP joint and transferred radially
Thumb and First Web Contracture

- Reconstruction of the Defect after Contracture release with local transposition or rotational flaps
- Index Transposition flap
- Dorsal Rotation flap
- Full Thickness Skin Graft from the Groin
In case of bony abnormality

- Radially based closing wedge corrective osteotomy of the Metacarpals, avoiding the Physes
- Thus Correcting the Ulnar Deviation of MC Head
<table>
<thead>
<tr>
<th>Cosmetic outcome</th>
<th>Count</th>
<th>Note</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>17 hands (13 hands were operated before 2 years)</td>
<td></td>
</tr>
<tr>
<td>Good</td>
<td>4 hands</td>
<td></td>
</tr>
<tr>
<td>Fair</td>
<td>2 hands</td>
<td></td>
</tr>
<tr>
<td>Poor</td>
<td>Nil</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Functional outcome</th>
<th>Count</th>
<th>Note</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>15 hands (14 hands were operated before 2 years)</td>
<td></td>
</tr>
<tr>
<td>Good</td>
<td>4 hands</td>
<td></td>
</tr>
<tr>
<td>Fair</td>
<td>3 hands</td>
<td></td>
</tr>
<tr>
<td>Poor</td>
<td>1 hand</td>
<td></td>
</tr>
</tbody>
</table>
Conclusion

- Early Surgery before 2 years of age
- Coupled with Long Term Dynamic Splinting
- Minimizing the Chances of Relapse