Dupuytren Disease
AAHS Review Course

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Case 1
Mother and daughter present same day

• Patient 1
  - 47 yr old
  - Age of onset
  - Associated conditions
  - Family history
  - Progression of disease
  - Risk factors

• Patient 2
  - 73 yr old
  - Onset 5 years ago

Diagnosis

• Initial findings
  - Nodules
  - Skin pit
  - Grapow fibers
  - More prominent volar web fat pad

• Differential Dx
  - Inclusion cyst
  - Ganglia
  - Giant cell
  - Tenosynovitis
  - Dermal lesions

Prevalence

• Varies widely based on population

• Hindocha 2009
  - 0.2 – 56%
  - 30% in Norwegians older than 60
  - 4% in English men over 40
  - 22% in Australians over 60
  - 1.8% in Japanese b/t 40-59
  - 56% in cross-sectional study of epileptics

Risk Factors

• Age
• Sex
  - More common in men
  - 9:1 decreasing to 2:1
• Family history
• Alcohol use
  - >15 / week
• ??? Association
  - Diabetes
  - Epilepsy
  - Smoking
  - Hand trauma

From R. Lanting 2014 – PhD Thesis
763 Netherland participants

Pathophysiology

• Contractile Myofibroblast
  - Differentiates from fibroblast
  - Found primarily in DD nodules
  - Contain organized bundles of alpha-smooth muscle actin (contraction)
  - Tightly bound to surrounding extracellular matrix and other cells
• Increased Type III > Type I collagen

Implicated genes involved with Wnt signaling
Case 2 - Pathoanatomy

- Palmar fascia
  - Longitudinal, transverse, vertical
- Bands – normal fascia
- Cords – pathologic

Pathoanatomy

- Palmodigital fascia
  - Grapow cord
  - Pretendonous cord
  - Central cord
  - Spiral cord
  - Lateral cord
- Radial aponeurosis
  - Commisural cords
- Ulnar aponeurosis
  - ADM cord

Spiral Cord

- Arises from
  - Pretendonous cord / proximal phalanx
  - Intrinsic muscle / ADM
  - Begin central and volar
  - Course dorsally and peripheral
  - Become superficial at PIP / P2 base

Ectopic Disease

- Garrod’s Nodes
  - Dorsal skin MP / IPs
- Lederhose Disease
  - Nodules plantar fascia
- Peyronie’s Disease
  - Plaque formation

- ADM Cord
- Commisural cord
**Clinical Stages of Dupuytren’s**

- **Early**
  - Proliferative Phase
  - Involucrin positive fibroblasts
  - Myofibroblasts
  - Nodule formation

- **Intermediate**
  - Residual Phase
  - Continued collagen deposition
  - Progressive contracture
  - Cord relative aneurysm

- **Advanced Disease**

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**Staging / Pattern**

<table>
<thead>
<tr>
<th>Tubiana Stage</th>
<th>Deformity</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No lesion</td>
</tr>
<tr>
<td>1</td>
<td>TFD between 0° and 45°</td>
</tr>
<tr>
<td>2</td>
<td>TFD between 45° and 90°</td>
</tr>
<tr>
<td>3</td>
<td>TFD between 90° and 135°</td>
</tr>
<tr>
<td>4</td>
<td>TFD greater than 135°</td>
</tr>
</tbody>
</table>

- Most common ring and small fingers – 80%
- Thumb disease – 3%

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**Indications**

- Hueston Table Top Test
- MP joints - contracture > 20 - 30°
  - High level success even in severe contracture
- PIP joints - contracture > 10-15°
  - Higher risk recurrent / residual contracture
  - Worse in higher severity
  - Worse in prolonged contracture

**Needle Aponeurotomy**

- Indications
  - Contracture with palpable cord
  - Cooperative patient
- Contra-indications
  - Scarring / adhered skin
  - Diffuse / nodular dz
- Technique
  - Identify portals
  - Distal to proximal
  - Anesthetize skin
  - 25 gauge 5/8” needle

**NA Aftercare**

- Cover with band-aids
- Elevate for 1-2 days
- No strenuous use / gripping activities 1-2 weeks
- No formal therapy
- Night splint PIPs
Efficacy / Recurrence

- Foucher et al. JHSb 2003
  - Initial improvement 76%
- van Rijsen JHSb 2006
  - Initial improvement 77%
- Recurrence (3.2 years)
  - Group of 100 hands
  - 59 hands needed further surgical Rx
  - Recurrence in 58 hands
- Recurrence (33 months)
  - 36 of 55 rays – 65%
  - 23 of 55 rays further Rx

Pess JHS 2012
Results of NA in 1013 fingers
minimum 3 year follow-up

- MP joints
  - Initial contracture – 35°
  - Correction <5°
  - 68% fingers
  - Reduction of contracture
    - 66% initial
    - 72% at final f/u
  - Recurrence
    - 20% of fingers
- PIP joints
  - Initial contracture – 50°
  - Correction <5°
  - 67% fingers
  - Reduction of contracture
    - 89% initial
    - 31% final f/u
  - Recurrence
    - 65% of fingers

Complications

- Minor complications
  - Skin tears
  - Swelling
  - Hematoma
  - Superficial infection
  - Paresthesias
- Major complications
  - Nerve injury: 0.05 - 2%
  - Flexor tendon laceration: 0.05%
  - Late flexor rupture
  - Arterial laceration

Five-Year Results of a Randomized Clinical Trial on Treatment in Dupuytren’s Disease: Percutaneous Needle Fasciectomy versus Limited Fasciectomy
Van Rijsen et al - PRS 2012

- 93 patients followed 5 years (84%)
  - Fasciectomy - 125 joints
  - NA – 167 joints
- Recurrence rates
  - NA group 85%
  - Fasciectomy group 21%
  - Recurrence earlier in NA
- Higher satisfaction fasciectomy group

Collagenase Clostridium histolyticum

- 64 yr old RHD
  - no family hx
- Left index PIP – 55°
  - Ulnar lateral cord
- Left small MP - 60° and PIP - 60°
  - Pretendonous cord
MOA

- Derived from Clostridium histolyticum
- 2 mixed Collagenase subtypes (AUX-I & AUX-II)
- More active than human collagenase
- Inactivation by local tissue substances
- Primary target = Fibrillar collagen (types I and III)

Administration

- Standard dose 0.58mg
- Up to 3 injections / cord
- 4 weeks between injections
- 1 or 2 injections / day

Injection

- Identify joint to be treated
  1. In fingers with combined contracture, typically treat MP before PIP joint
- Identify optimal location along cord for injection
  1. Maximal separation of cord from flexor sheath
  2. Area of cord not adhered to overlying skin
- Reconstitute 0.58 mg of clostridial collagenase in 0.25ml (MP) or 0.20ml (PIP) diluent
- Inject dose into 3 contiguous areas of cord
  1. Avoid extravasation - should feel significant resistance

Manipulation

- Up to 72 hours after injection
  1. If contracture persists, perform finger manipulation
  2. Typically performed with local anesthesia
- Maximal extension splint made to be worn at night for 1 – 4 months
- No formal therapy – instructed in edema control and range of motion
- Avoid forceful grasping for up to 4 weeks

Reduction of contractures to <5° after up to 3 injections

- Randomized, double-blind, placebo-controlled
  (n=306)

Contrastive Bar Chart

- PIP joint n=70
- MP joint n=133
- All n=203
### 3 Year Recurrence Data

![Graph showing recurrence data]

- MP-Low: Baseline contracture ≤50°
- PIP-Low: Baseline contracture ≤40°

#### Key Points
- Successfully treated joints ≤5°
- MP-Low: Baseline contracture
  - 28%
- PIP-Low: Baseline contracture
  - 18%
- 50%
- 71%

### Complications

- Common complications (≥30%)
  - Bruising
  - Swelling
  - Injection site tenderness
  - Skin blister
- Other minor complications (≥5%)
  - Pruritis
  - Lymphadenopathy / lymph node pain
  - Skin tear
- Treatment
  - Elevation, compression
  - Antihistamines, analgesics
  - Wound care

### Day of Manipulation vs. 12 Days post

- **Skin Tears:** 10-15%
  - Heal primarily
  - Local wound care
  - Higher risk
    - Multiple injection
    - Severe contractures
    - Areas of scar / skin adherence
- **Serious Complications**
  - Tendon Rupture
    - Clinical trials
      - Rupture in 3 out of 1082 patients
      - Onset average 6 days (4, 6, 8) after injection
      - All followed small finger injection for PIP contracture
        - FDS and FDP (3)
        - FDP and partial FDS (1)
    - 3 year safety data
      - 49, 078 injections
      - 26 tendon ruptures (0.05%)
- **Allergic Reactions**
  - Exposure to bacterial collagenase triggers antibody response
  - 30 days after 1st injection
  - 92% patients +Ab to Aux-1
  - 91% patients +Ab to Aux-2
  - 100% patients seropositive after 4th injection
  - Incidence of pruritis ↑ with # injections
- **Anaphylaxis**
  - One reported case at this time
  - MDs should be prepared to treat
    - Epinephrine 0.3mg SQ/IM q5 min prn
    - IV with NS 5-10 ml/Kg in 1st 5 min
    - Diphenhydramine 25-50 mg q2-4 hr prn
- **Pulley Rupture**
  - A2 and A4
  - Following small finger injection for PIP contracture
- **Tendonitis**
- **CRPS**

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**Surgical Management**

- **Fasciotomy** (open or percutaneous)
  - Disruption of cord
- **Fasciectomy**
  - Removal of diseased fascia
- **Dermofasciectomy**
  - Excision of cord and skin
  - Wound covered with skin graft

**Post-Op Care**

**Rehabilitation**

- **Goals**
  - Maintain maximal extension
  - Regain full flexion
  - Speed time to normal use
  - Limit scar contracture

- **Static splint made to maintain maximal extension**
  - Worn at night for 3 – 6 months
  - Interval use during day until regain comfortable AROM

- **Daily exercises**
  - Isolated active PIP, DIP and composite motion
  - Edema control
  - Scar remodeling
  - Strengthening

- **Other interventions**
  - Dynamic flexion or extension splints

**Subtotal Palmar Fasciectomy**

- Anesthesia may be local or regional
  - Location of abnormal tissue
  - Surgeon preference

- Identification of diseased tissue

**Dissection of the involved tissue**

- Careful identification of digital nerves and arteries

- Isolation and removal of cord(s)
Recurrence after surgery
- 19% - 54% at 5 years

Becker et al. 2010
- 69 articles on surgery for primary Dupuytren's between 1946 and 2009
  - 57 retrospective case series
  - 7 prospective case series
  - 2 prospective randomized controlled trials
  - 3 prospective randomized controlled clinical trials
  - 51 papers reported rates of recurrence, ranging from 0 to 71%

Becker et al. The outcome of surgical treatments from primary Dupuytren's disease--a systematic review. The JHS 2006;57:7–13

Complications
- Most common
  - Nodule formation
  - Cord formation
  - Nodule formation in 4%
  - Cord formation in 8%

- 7% under grafted area
- 39% outside of grafted area

- Progressive fibroproliferative disorder
  - Underlying causes not yet elucidated
  - No known way to alter the course of disease

- Recruitment – need a prospective comparative study
  - Least with fasciectomy
  - Highest with Needle aponeurotomy

- Return to use
  - Quickest with Needle and collagenase
  - Longest with fasciectomy

- Complications
  - Serious complications uncommon with all options
  - Discuss thoroughly with patient and let them participate in choosing best treatment for them

Summary
- Complication rates range from 15% - 36%
- Most common
  - Nodules
  - Skin necrosis

- Risk of higher complication rate
  - Primary surgery with MP contracture > 30° (Bulstrode 2005)
  - All digital nerve or artery lacerations in this group
  - Secondary surgery for recurrent contracture (Coert 2006)
  - 3% incidence of nerve injury in primary surgeries vs 12% in secondary surgeries.