Importance of Dupuytren severity assessment

- Guide treatment recommendations
- Compare treatment outcomes

No universal Dupuytren classification system exists (Akhavani 2015)

What does severity mean to you?

- **Primary anatomy**
  - Is fibrosis diffuse or localized?
  - How many fingers/joints are involved?
  - How involved is the skin?
  - How long does surgery take?
  - How likely is a major (nerve / artery / tendon) complication?
  - How successful is intraoperative correction of contracture?

- **Secondary pathoanatomy**
  - PIP capsule contracture
  - Flexor sheath contracture
  - Central slip compromise / boutonniere
  - Sagittal band rupture

- **Biological:**
  - What is the contracture/recontracture velocity?
  - How active (nodular) is the local disease?
  - How likely is excessive/prolonged postop inflammation (flare)?

- **Patient reported quality of life impact**

Actuation, Progression, Correction, Recontraction, Stabilization

- **Activation:** Risk of ever developing Dupuytren-like findings
  - Demographic
    - ↑↑ if Positive family history (Hindocha 2004, Becker 2014)
      +FHx ↑ likelihood early onset, ectopic disease, high severity score (below)
    - ↑ if Caucasian (In US Caucasian > Hispanic > Black > Asian) (Saboeiro 2000)
    - ↑ if Greater age (Lanting 2014, Descatha 2014)
    - ↑ if Male gender (Lanting 2014)
    - ↑ if high Genetic score (Dolmans 2011, Dolmans 2012)
      (WNT4 WNT7B WNT7B WNT2 RSPO2 MAFB DMRT1 DMRT1 EPDR1)
  - 10 X more DD patients without contracture than with (Diep 2015)
  - Less gender difference in asymptomatic disease (Diep 2015)
  - Dupuytren Spectrum diseases effect on risk of developing Dupuytren
    - ↑ if Ledderhose (Schurter 2017)
    - ↑ if Frozen Shoulder (Smith 2001), esp. women (Degref 2013)
    - ± if Peyronie (Data sampling problem: demographic overlap)
  - Mechanical
    - ↑ if Acute trauma (Abe 2007, Rayan 2005)
Indicators of Dupuytren Biologic Severity

- if Hand-transmitted vibration exposure (Palmer 2014, Descatha 2014)
- if Rock Climbing (Logan 2005)

- Comorbidities: Effect of conditions on risk of developing Dupuytren:
  - if Diabetes (Descatha 2014)
  - if Psoriasis (Patel 2014)
  - if Underweight (Gudmundsson 2000)
  - if Hyperlipidemia (Sanderson 1992)
  - if Joint stiffness (Williams 2015)
  - if Smoke: dose-dependent (Godtfredsen 2004) but \textit{only if} (-)FHx (Becker 2014)
  - if Heavy drinking (Godtfredsen 2004, Descatha 2014)
  - if Combination heavy smoking and heavy drinking (Godtfredsen 2004)
  - if Overweight (Hacquebord 2016)
  - if Rheumatoid arthritis (Arafa 1984)

- **Progression**: risk (not rate) of progressing from nodule to contracture
  - Data (mostly) supports relationship:
    - if + Family History: dose response (Becker 2014)
    - if Current smoking: dose response (Eckerdal 2014, Descatha 2014)
    - if Daily alcohol: dose response (Descatha 2014); also present with more severe contractures (but duration not known: do they wait longer?) (Hindocha 2008)
    - if Years using vibrating hand tools: dose dependent (Descatha 2014)
    - with \uparrow surface area of involvement (Lanting 2016)
  - Pseudo-Dupuytren: early cords/nodules \textit{without} progression
    - Post injury / postop (Beasley 2003, Abe 2007, Rayan 2005)
  - Not clear one way or the other impact on risk of progressive contracture
    - ± Epilepsy (Strong pro and con data - both epilepsy and antiepileptics)
    - ± Peyronie (No data)
    - ± Frozen shoulder (No data)

- **Correction**: What’s the expected early outcome? Exam more influence than diathesis
  - McFarlane outcome formula to predict early outcome (Legge 1980)
    - Worse with \uparrow \# involved rays, small finger procedure, PIP, \uparrow Preop contracture
  - Factors predicting lack of full correction (Adam 1992)
    - Worse: PIP joint, Redo PIP, small finger PIP, more than one ray involved
    - \textit{Not} age, gender, occupation, alcohol
  - PIP release worse than only manipulation esp. if pinned (Breed 1996)
  - Incomplete correction more likely for MCP > 50, PIP >40 (Withthaut 2013, Schulze 2014, Verheyden 2014)
  - Procedure: PIP correction more likely with surgery than CCH (Zhou 2016)
  - Joint: PIP correction less likely than MCP correction (Badalamente 2013)

- **Recontraction**: Rate of post-treatment recontraction
  - \textit{True vs false recurrence} (Dias 2013, Eaton 2015)
    - Early: 1st 6 weeks then plateau: persistent secondary pathoanatomy
    - Progressive: no plateau: persistent biologic activity
    - Late: after a year or more of stability: true recurrence (reactivation)
Diathesis factors
- Predict relative risk of recontraction
  - Don’t clearly predict early complications or short term outcome
    - Hueston’s original: four factors (Hueston 1963)
      - Caucasian, bilateral, any family history, any ectopic disease
    - Hindocha score: five factors (only Caucasians studied) (Hindocha 2006B)
      - Bilateral, parent/sibling family history, knuckle pads, male gender, diagnosis age 50 or younger
      - Each factor increases risk; having all 5 triples risk compared to none
    - Abe Diathesis score (Abe 2004)
      - 1 point each for bilateral, small finger surgery, onset < 50 y
      - 2 points each for Ledderhose, knuckle pads, thumb / index disease
      - Diathesis score = sum of points. Increased risk for score > 4.
  - Degreef Risk Factors for recurrence (Degreef 2011)
    - Bilateral, ectopic disease, onset < 50 y, > 2 two rays affected, thumb disease, + family history, small finger surgery, male
    - High genetic risk score associated with age of onset younger than 50, positive family history, knuckle pads, and Ledderhose (Dolmans 2012)
- Age < 60 at time of surgery (Rombouts 1989)
- Histology of excised tissues
  - Risk: Proliferative > Fibrocellular > Fibrotic (Rombouts 1989)
  - Recurrence rate of proliferative 3X Fibrotic (Balaguer 2009)
  - Independent of Diathesis factors (!) (Balaguer 2009)
  - Cords of more severe contractures less cellular (Verjee 2009)
  - The rock and the hard place:
    - Less severe contractures: more biologically active; ↑ recurrence
    - More severe contractures: less biologically active; ↑ 2° pathoanatomy
- Procedure type: Recurrence rate after minimal twice that of fasciectomy
- Success: lower recontraction rate with full correction (Dias 2006, Peimer 2013)
- Angle: higher recurrence rate for pretreatment PIP > 40° (Peimer 2013), MCP > 50° (Peimer 2015)
- Joint treated: PIP much more rapid than MCP; isolated PIP worse (Crowley 1999)
- Finger treated: Greater risk small finger (Degreef 2011)
- No clear effect on recurrence: (Hindocha 2006B, Degreef 2011)
  - Smoking, Alcohol, Frozen shoulder, Diabetes, Manual labor, Prior injury
- Stabilization, regression (Reilly 2005, Lanting 2016) and stress shielding (Verjee 2009, Melamed 2017)
  - If minimal contracture: Pseudo-Dupuytren
  - If moderate contracture: stress shielding from joint capsule contracture
  - If severe contracture: stress shielding from disuse / functional amputation

Most influential factors
- Activation: Parent / sibling with disease
- Progression: Family history, prior contracture
Correction: Contracture location / angle
- Early Loss of Correction: Residual anatomic factors
- Progressive Recontraction: Diathesis, procedure
- Prolonged Stabilization: Diathesis, stress shielding

Additional Points

- Recontracture is a rate over time, not a fixed risk
- “Average follow up” duration spanning multiple years is bad data
- Subset of patients requiring procedures: selection bias - more severe
- Subset of patients achieving full correction: selection bias – lower recurrence
- Almost every Dupuytren association is disputed somewhere

Severity Score (Hindocha 2006A) correlates independently with positive family history

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Score</th>
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<tbody>
<tr>
<td># DC procedures</td>
<td>Total both hands</td>
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<tr>
<td># Individual digit recurrences</td>
<td>Total all digits</td>
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<tr>
<td># Digits affected</td>
<td>Total all digits</td>
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<tr>
<td># Nodules</td>
<td>Total both hands</td>
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<tr>
<td># Pits</td>
<td>Total both hands</td>
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<td>Garrod Pads</td>
<td>Present = 1</td>
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<tr>
<td>Ledderhose</td>
<td>Present = 1</td>
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<td>Total Tubiana score</td>
<td>Sum of all joint contactures / 45’</td>
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<tr>
<td># hands affected</td>
<td>1 or 2</td>
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<tr>
<td>Total Severity Score</td>
<td>Sum of above</td>
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</tbody>
</table>

Pros: Doesn’t rely on patient’s memory

Cons: Cumbersome; no time stamp to assess rate of progression
### Summary: Published Factors Influencing Risk of Dupuytren Events

<table>
<thead>
<tr>
<th>Events</th>
<th>Factors</th>
<th>Diagnosis</th>
<th>Progression</th>
<th>Partial correction</th>
<th>Recontracture Rate</th>
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<tbody>
<tr>
<td>↑ Increase</td>
<td>↑ Decrease</td>
<td>--- No effect</td>
<td>± Contradictory data</td>
<td>[blank] No data</td>
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<tr>
<td>Family History</td>
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<td>↑ Current age</td>
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<td>Frozen Shoulder</td>
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<td>Smoking</td>
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<td>Excess Alcohol</td>
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<td>Cm² area involved</td>
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<td>Full correction</td>
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<td>↓ Age at treatment</td>
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<td>Thumb involvement</td>
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<td>CCH / PNF</td>
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<td>Open PIP release</td>
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<td>Proliferative histology</td>
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<td>Fibrotic histology</td>
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More materials at [https://dupuytrens.org/2017-ash-symposium-7/](https://dupuytrens.org/2017-ash-symposium-7/)
References


Indicators of Dupuytren Biologic Severity

Charles Eaton MD


