Esophageal Strictures

Where are we and where do we need to go?

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The Hospital for Sick Children
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Objectives

• Review incidence and clinical characteristics of esophageal strictures

• Literature

• EBCRC data

• Toronto led multicenter retrospective study

• Review management controversies
Esophageal Strictures

- How common are they?
- Who is at risk?
- Where are they located?
# Esophageal Strictures

## How common are they?

<table>
<thead>
<tr>
<th>Source</th>
<th>n/N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>JD Fine, JPGN 2008; 46: 147-58</td>
<td>254/2627</td>
<td>10</td>
</tr>
<tr>
<td>Freeman EB, BJD 2008; 158: 1308-14</td>
<td>39/223</td>
<td>17.4</td>
</tr>
<tr>
<td>EBCRC database</td>
<td>90/692</td>
<td>13</td>
</tr>
</tbody>
</table>
## Esophageal Strictures

### Who is at risk?

#### EB subtype

<table>
<thead>
<tr>
<th>Source</th>
<th>JEB-H</th>
<th>JEB-nH</th>
<th>DDEB</th>
<th>RDEB loc</th>
<th>RDEB gen</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>JD Fine</strong>&lt;br&gt;<strong>JPGN 2008; 46: 147-58</strong>&lt;br&gt;(n=254)</td>
<td>14</td>
<td>30</td>
<td>4</td>
<td>37</td>
<td>80</td>
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<tr>
<td><strong>Freeman EB BJD 2008; 158: 1308-14</strong>&lt;br&gt;(n=223)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>65</td>
</tr>
<tr>
<td><strong>EBCRC database</strong>&lt;br&gt;(n=226)</td>
<td>0</td>
<td>10</td>
<td>10</td>
<td>-</td>
<td>60</td>
</tr>
</tbody>
</table>
Esophageal Strictures

Who is at risk?

EB subtype

Toronto Study (n=63): 98% DEB
- RDEB gen intermediate: 27%
- RDEB generalized severe: 54%
- DDEB generalized: 4.2%
- Other DEB subtypes: 14.6%

81%
Esophageal Strictures

Who is at risk?

Age

FIG. 1. Cumulative risk of esophageal strictures and stenoses in inherited epidermolysis bullosa. EB = epidermolysis bullosa; S = simplex; WC = Weber-Cockayne variant; K = Koebner variant; O = all other variants; J = junctional; H = Herlitz variant; nH = non-Herlitz variants; DD = dominant dystrophic; R = recessive; HS = Happleau-Siemens variant; nHS = non-Happleau-Siemens variant; I = inverse variant.
Where are they located?

• ~75% upper and middle esophagus
• Single stricture > multiple
• Short segment (2-5 cm)


JD Fine JPGN 2008; 46: 147-58
Freeman EB BJD 2008; 158: 1308-14
Management
Prevention

Primary

- Feeding modifications
- Antireflux medication

Secondary

- Steroids
- Mitomycin C
Primary Prevention

- No evidence
- Monitoring in high risk groups
- Antireflux medication
Secondary Prevention

**Peri-procedural**

- **Oral dexamethasone**
  - 1-2 mg/kg during procedure, wean after 5 days
- **Mitomycin C**
  - Antifibrotic, antiproliferative
  - 0.1mg/ml for 2-3 mins
- **Budesonide oral viscous sol**
  - 16 patients

**Post- procedural**

- **Budesonide oral viscous sol**
  - 0.5 mg/2ml budesonide capsule mixed with 5 gm sucralse and maltodextrin
  - OD/BID
  - Longest duration: 18 months
  - Decreased number of dilatations
  - ? Increased yeast infection
  - ? Adrenal suppression

References:

Azizkhan R. J Ped Surg 2006; 41:55-60
Endoscopy 2016;48(01):71-74

TrossenenS. JPGN 2007;44:336-41
Dohil R. JPGN 2011;52(6):776-7
Management of Dilatation

- Antegrade vs Retrograde Approach
- Fluoroscopy vs Endoscopy
- Sedation vs General Anesthesia
- Adjuvant medical treatment
Management of Dilatation

Antegrad Approach

Pros:
• Lower aspiration risk
Cons:
• Mouth trauma
• Need for GA
• More difficult for proximal strictures

Retrograde Approach

Pros:
• Less mouth trauma
• Better for proximal lesions
• Sedation
Cons:
• Need for G-tube
• Increased aspiration risk

Azizkhan R. J Ped Surg 2006; 41:55-60
Spiliopoulos S AmJ Roetgen 2012;199:208-12
Goll G Diseases esoph 2017;30:1-6
De Angelis P. J Ped Sx 2011;46:842-7
Castillo R. JPGN 2002;34):535-41
Anderson S. GI Endosc 2004; 59:28-32
Management of Dilatation

Fluoroscopy

Pros:
• Lower perforation risk
• Antegrade and retrograde approach
• Less mouth trauma

Cons:
• Radiation risk

Endoscopy

Pros:
• Direct visualization

Cons:
• Increased risk of perforation
• Need for GA

References:
Azizkhan R. J Ped Surg 2006; 41:55-60
Spiliopoulos S AmJ Roetgen 2012;199:208-12
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Management of Dilatation

Sedation

Pros:
• Quicker recovery
• Less blistering

Cons:
• Aspiration risk
• Emergency airway may be extremely difficult
• Hypoventilation

General Anesthesia

Pros:
• Lower perforation risk

Cons:
• Increased blistering
• Longer recovery
• Increased endotracheal scarring

Gottschalk A. Curr Opin Anesthesia 2010; 23:2(%);18-22
Gollu G. Dis Esophagus 2017;30:1-6
Management of Dilatation

- All studies report > 95% success rate
- Repeat procedures more related to underlying disease rather than procedure
- Higher risk of perforation with endoscopy
Toronto Preliminary Data

- Fluoroscopy 63%
- Endoscopy 22%
- Retrograde approach 10%
- Bougienage - 3%
- Unspecified - 2%

63 patients
136 strictures
99 dilatations
Management of Dilatation

Adjuvant medical treatment

• Steroids

• Mitomycin C

• Antibiotics

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Endoscopy 2016;48(01):71-74

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Toronto Preliminary Data

- PPI 50%
- Systemic steroids 41%
  - Budesonide - 10%
  - Phenytoin - 5%
- Others, unspecified - 24%

63 patients
136 strictures
99 dilatations
68% meds
Management of Dilatation

• There is no consensus on the best approach
• Considerations:
  • Team’s expertise
  • Location and length of the strictures
  • Presence of G-tube
  • Recurrences
  • ? role of medical treatment
Summary

• Risk factors of esophageal strictures depend on the age of the patient and EB subtype
• There is no consensus on the best approach for the dilatation
• There is no evidence that medical treatment may prevent strictures
• The use of budesonide slurry should be further explored
• Need for consensus guidelines
EB patients/families

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Dr. Julio Salas
Dr. Ignacia Fuentes
Dr. Anna Martinez
Dr. Jemima Mellerio
Dr. Dedee Murrell

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EB & MRF