

# Esophageal Strictures

*Where are we and where do we need to go?*

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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?

Source	n/N	%
<i>JD Fine</i> <i>JPGN 2008; 46: 147-58</i>	254/2627	<b>10</b>
<i>Freeman EB</i> <i>BJD 2008; 158: 1308-14</i>	39/223	<b>17.4</b>
<i>EBCRC database</i>	90/692	<b>13</b>

# Esophageal Strictures

Who is at risk?

EB subtype

Source	JEB-H	JEB-nH	DDEB	RDEB loc	RDEB gen
<i>JD Fine JPGN 2008; 46: 147-58 (n=254)</i>	14	30	4	37	80
<i>Freeman EB BJD 2008; 158: 1308-14 (n=223)</i>	0	0	0	-	65
<i>EBCRC database (n=226)</i>	0	10	10	-	60

# Esophageal Strictures

Who is at risk?

## EB subtype

**Toronto Study (n=63): 98% DEB**

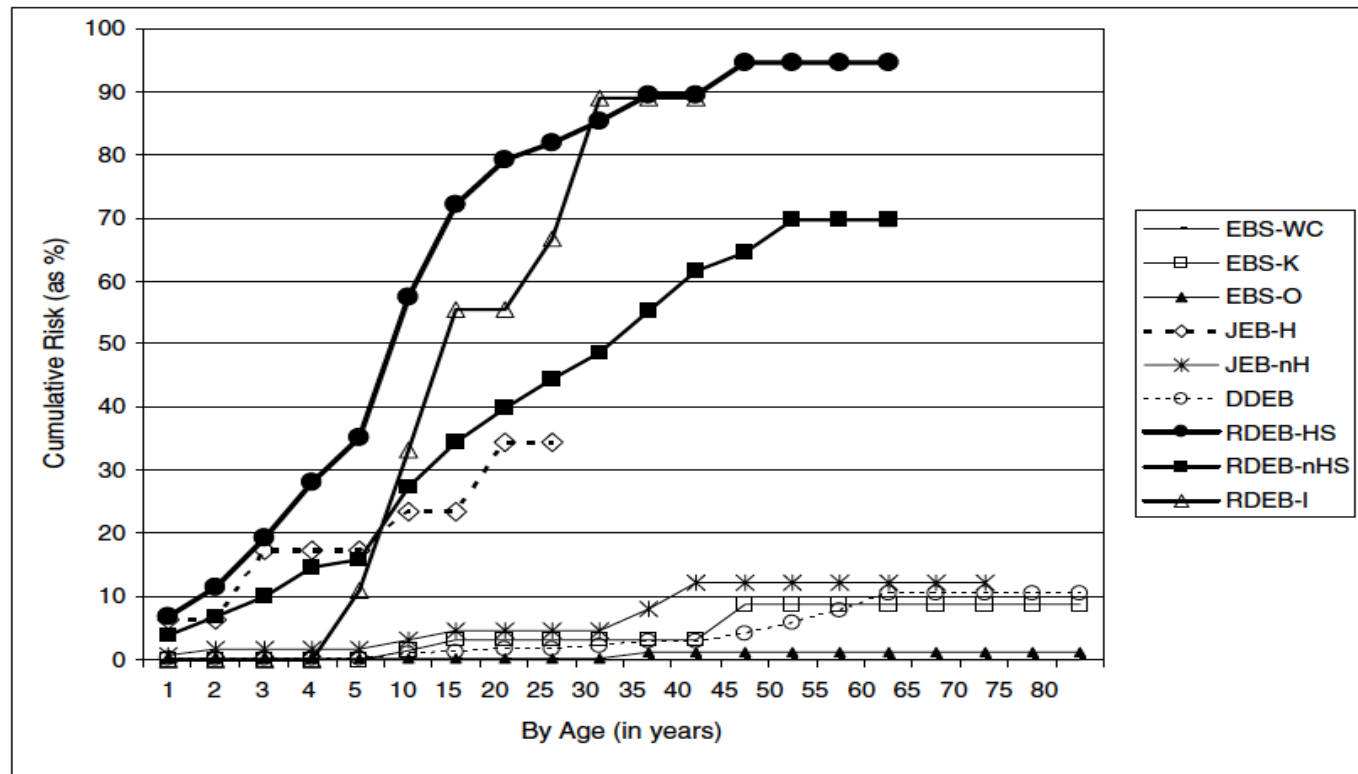
- RDEB gen intermediate: 27%
- RDEB generalized severe: 54%
- DDEB generalized: 4.2%
- Other DEB subtypes: 14.6%

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 = Hallopeau-Siemens variant; nHS = non-Hallopeau-Siemens variant; I = inverse variant.

# Esophageal Strictures

## 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*

*Rev. Bras.*  
*Otorrinolaringol. vol.74 no.5 São*  
*Paulo Sept./Oct. 2008*



# Management



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# 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
  - 16 patients
- Budesonide oral viscous sol

## Post- procedural

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

*Azizkhan R. J Ped Surg 2006; 41:55-60*  
*El-Asmar KM J Ped Surg 2013; 48(6):1454-7*  
*Endoscopy 2016;48(01):71-74*

*TrossenenS. JPGN 2007;44:336-41*  
*Dohil R. JPGN 2011;52(6):776-7*  
*Zanini A. Ped Drugs 2014;16:391-5*

# Management of Dilatation

**Antegrade vs Retrograde Approach**

**Fluoroscopy vs Endoscopy**

**Sedation vs General Anesthesia**

**Adjuvant medical treatment**

# Management of Dilatation

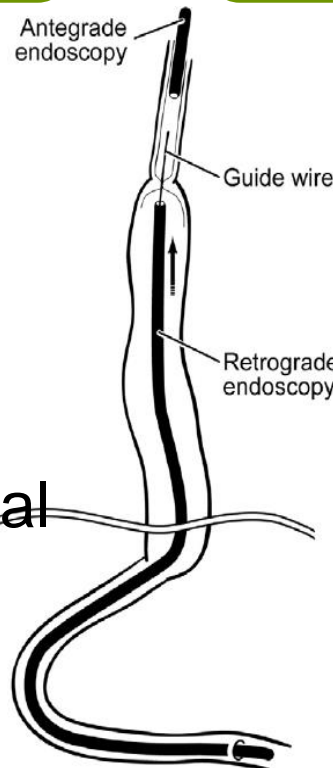
## Antegrade Approach

### 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

*Azizkhan R. J Ped Surg 2006; 41:55-60  
Spiliopoulos S AmJ Roetgen 2012;199:208-12  
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Castillo R. JPGN 2002;34):535-41  
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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*

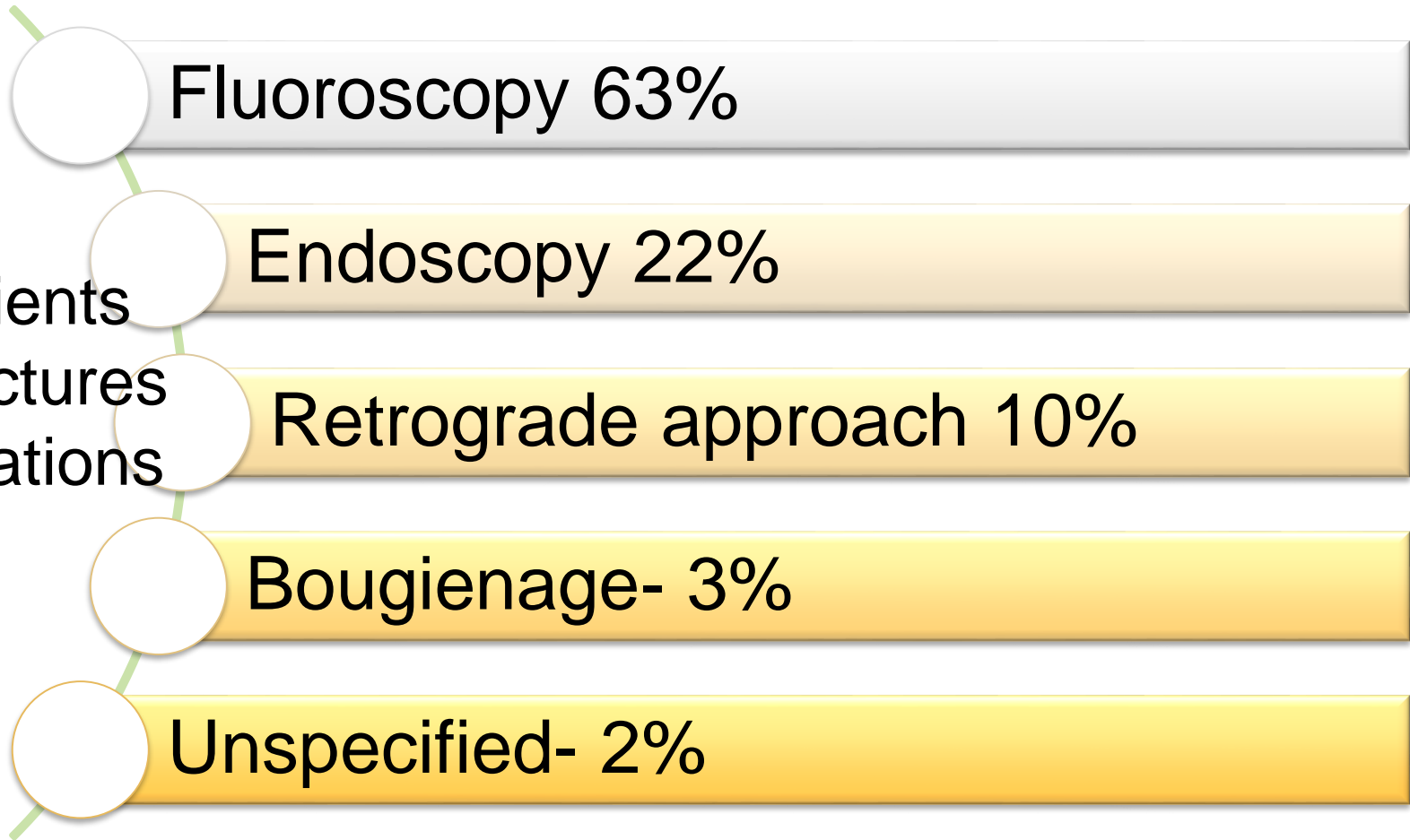


# Management of Dilatation

- 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

63 patients  
136 strictures  
99 dilatations



# Management of Dilatation

## Adjuvant medical treatment

- Steroids
- Mitomycin C
- Antibiotics

*Azizkhan R. J Ped Surg 2006; 41:55-60*  
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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

## Collaborators

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Dr. Anna Martinez

Dr. Jemima Mellerio

Dr. Dedee Murrell



## EBCRC-THE EPIDERMOLYSIS BULLOSA CLINICAL RESEARCH CONSORTIUM



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