





Dysphagia after EA repair

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Disclosure

In the past 12 months, I have had no relevant financial relationships with the manufacturer(s) of any commercial product(s) and/or provider(s) of commercial services discussed in this CME activity.



Learning objectives

- Incidence of dysphagia after EA repair
- Differential diagnosis: causes and mechanisms of dysphagia after EA
- How to practically manage a children presenting with dysphagia after EA



Esophageal Atresia

- Most frequent upper GI malformation
- Incidence : 1:2,500 to 3,000

8%
1%
84%
3%
4%

Incidence and symptoms of dysphagia after EA repair

- Incidence of dysphagia ranges from 39 to 77%
- May occur early in life or later.

Early	Late
Regurgitations	Issue to swallow solid food
Difficulties to introduce solid food	Food impaction
Food refusal	Slow eater
Aspiration, Failure to thrive	Need to drink during meals
	Vomiting
	No reported complaint... But actual symptoms!!!

Chetcuti et al. Arch Dis Child 1993;68:163-6.
 Little et al. J Pediatr Surg 2003;38:852-6.
 Somppi et al. J Pediatr Surg 1998;33:1341-6

Upper GI Symptoms in EA

40 patients, 11 months – 18 years;
 EA type A n=5 type C=35

18/40 = 45% w/dysphagia

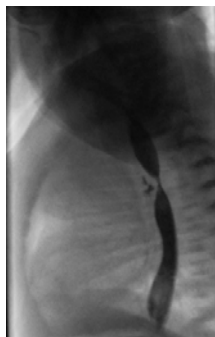
Symptoms	All patients (n=40)
GERD Symptoms	
Asymptomatic (no complaint)	7 (18%)
Pyrosis	6 (15%)
Regurgitation	8 (21%)
Heartburn	6 (15%)
Nausea	7 (18%)
Vomiting	5 (13%)
Obstructive Symptoms	
Blockage/Impaction	12 (32%)
Liquids	1 (3%)
Thickened Liquids	2 (6%)
Soft Foods	5 (13%)
Dry Foods	11 (29%)
Solids	14 (37%)
Change in Alimentary Habits	
Need to drink	17 (45%)
Change in diet	20 (53%)
Last to finish meal	13 (34%)

Lemoine et al IPGN 2013

Case #1

- Clementine, 1 ½ month
- Born 39 weeks, EA type C
- Thoracoscopy, anastomosis at day 1 with tension
- Discharge from hospital at day 12 with PO lansoprazole
- Seen at day 45 for regurgitations+++
- Baryum swallow

Anastomotic stricture



Anastomotic stricture

Anastomotic strictures that require dilatation develop in 24 to 79%

Study, year	Subjects, no. of type C	Stricture rate	Definition of stricture
Chittmittrapap et al 1990 ²³	N = 184 (not reported)	37% (74)	Required dilatation
Poeraru et al 1991 ²⁰	N = 74 (74)	24% (18)	Required dilatation
Engum et al 1995 ²	N = 215 (178)	35% (75)	Required dilatation
Konkin et al 2003 ¹⁶	N = 136 (119)	52% (69)	Not stated
Lain et al 2007 ⁴	N = 34 (29)	79% (27)	Required dilatation
Serhal et al 2010 ¹⁹	N = 64 (64)	37% (23)	Contrast esophagram
Alshehri et al 2012 ³	N = 50 (39)	36% (18)	Required dilatation
Koivusalo et al 2013 ⁴	N = 127 (110)	78% (102) 38% > 5 dilatations	Based on endoscopy
Total	884	40.0% (353)	



Anastomotic stricture

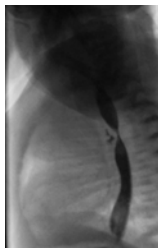
- Predisposing factors are anastomotic tension, anastomotic leakage, and presence of GER
- Previous studies have shown a clear correlation between GER and esophageal strictures secondary to EA/TEF, with an incidence as high as 60%



Eur J Ped Surg 2012



Anastomotic stricture

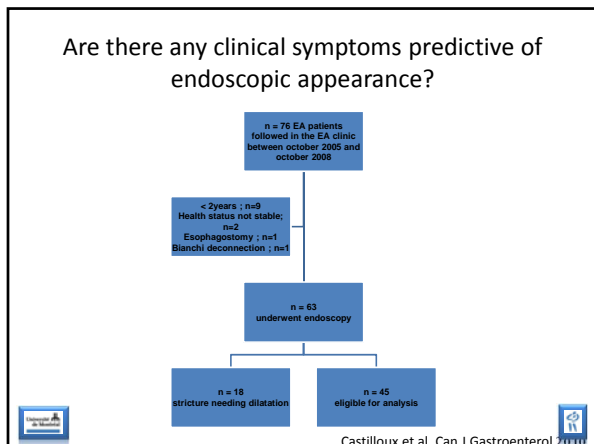


Gastroesophageal Reflux & Peptic Esophagitis

- Reflux is almost universal after EA repair
- About 50% of patients may be weaned of antireflux treatment
- Symptoms are non specific
- Long term complications include esophagitis, Barrett esophagus, esophageal cancer

Okada et al. JPS 1997
Koivusalo et al. JPS 2007
Tovar Dis Esophagus 2013



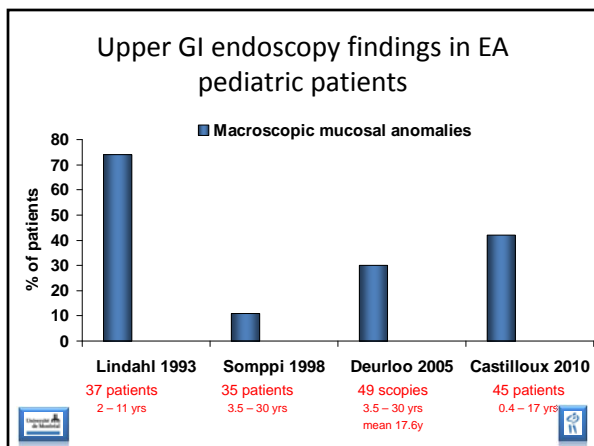


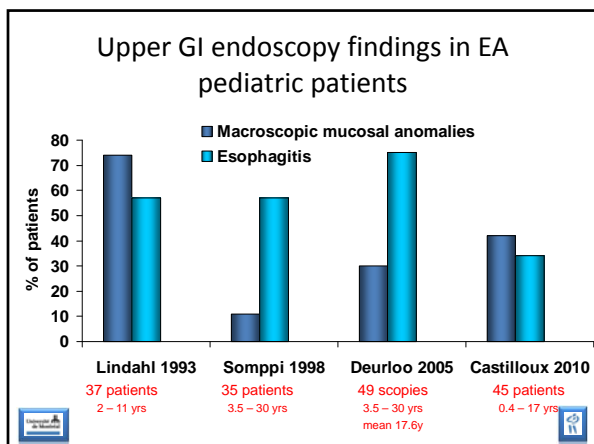
Symptoms vs. Endoscopy

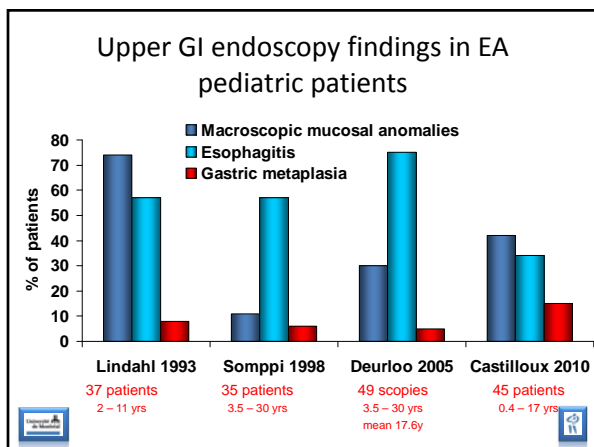
	Normal endoscopy n= 26	Abnormal endoscopy n = 19
Regurgitation / clinical GER	15 %	26 %
Pyrosis	8 %	21 %
Dysphagia	38 %	42 %
Odynophagia	4 %	0 %
Food impaction	38 %	32 %
Cough at meals	23 %	32 %
Asymptomatic	42 %	32 %

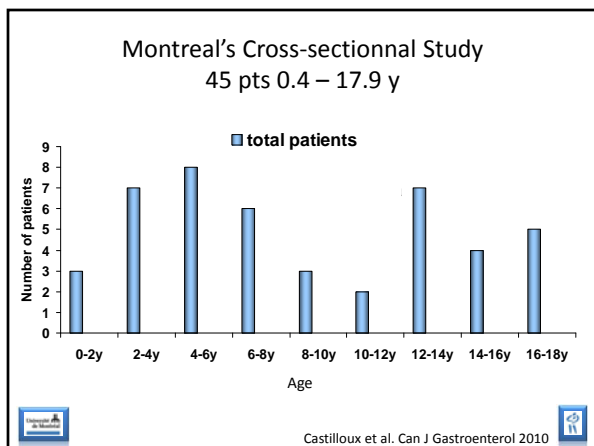
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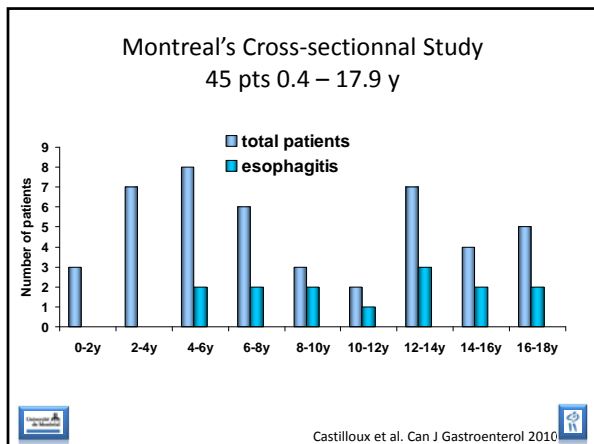
Castilloux et al. Can J Gastroenterol 2010

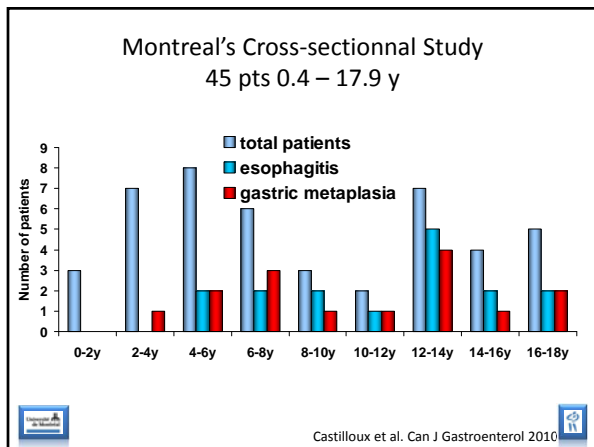










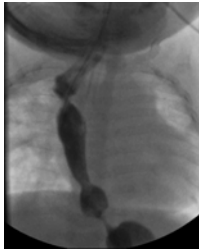


Congenital Esophageal Stenosis

- Although CES is rare (1:50,000) in the general pediatric population, its association with EA is frequent ranging between 8% and 14% in the largest series
- Diagnosis is difficult : mild CES can be interpreted as transient spasm or dysmotility

Newman B, Bender TM. *Pediatr Radiol* 1997;27:530-4.
Kawahara Het al. *Surgery* 2001;129:29-38.
Yoo H et al. *Pediatr Radiol* 2010;40:1353-1359.

Boy 9 months, EA type A, VACTERL.



Anastomotic stricture and congenital stenosis



Esophagoscopy showing the CES before (right) and after (left) pneumatic dilation

Girl, EA type C, Thoracoscopy

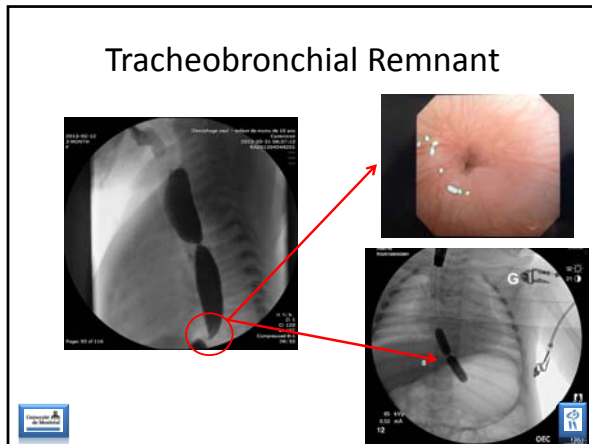


Post Op 15 opacification

2 ½ months : Regurgitations, Desaturation



Baryum swallow, Day 49



Congenital Esophageal Stenosis

- CES associated with EA is frequent.
- A high suspicion index for CES must remain in the presence of EA.
- It can be diagnosed at the time of EA repair or on the first postoperative esophagogram.
- Dilatation may be effective to treat some of them but perforation is frequent.

McCann et al Dis Esophagus 2013

University of Michigan

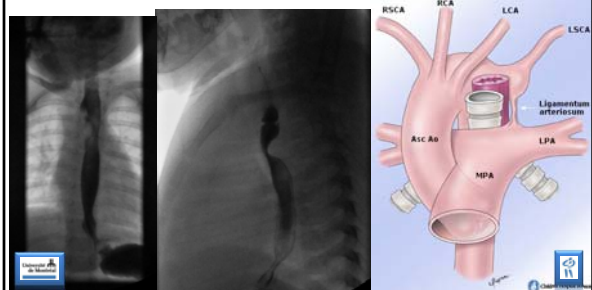
Vascular malformations

- Occur in 2-5% of EA
 - Vascular ring
 - Aberrant right subclavian artery
- Compress esophageal wall and may impair esophageal emptying

University of Michigan

Vascular Ring

- Right Aortic Arch, aberrant left subclavian artery & anterior ligamentum arteriosum

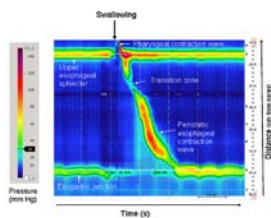


Aberrant right subclavian artery

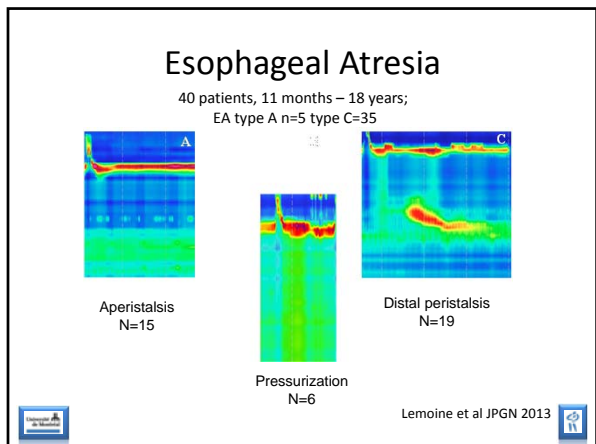


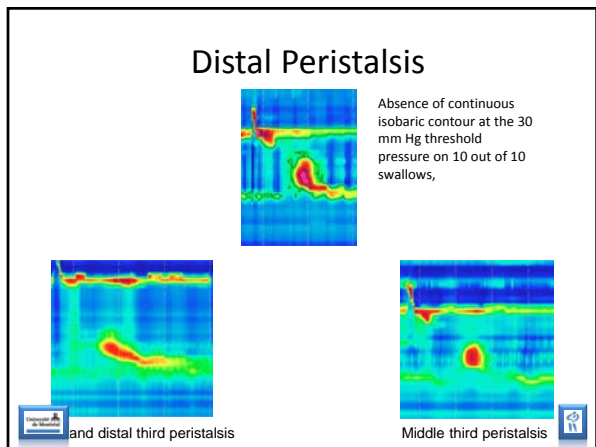
Dysmotility

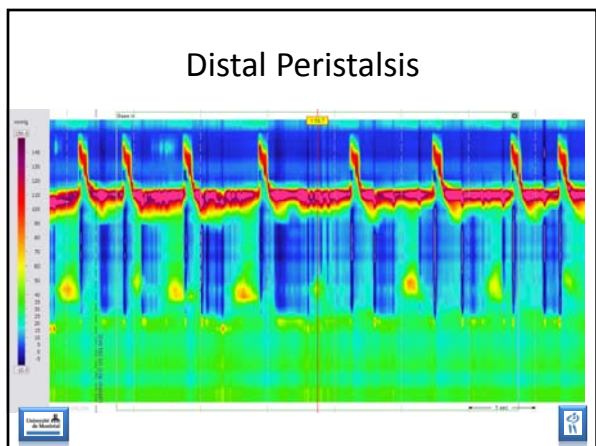
- All patients with EA have an abnormal esophageal motility



Lemoine et al JPGN 2013







Motility vs. Symptoms

Symptoms	Peristalsis Classification				
	Aperistalsis (n=15)	Pressurisation (n=6)	Distal Peristalsis (n=19)		
Asymptomatic (no complaint)	4 (26%)	1 (17%)	2 (11%)		
GERD Symptoms	Pyrosis	3 (23%)	0 (0%)		
	Regurgitation	4 (31%)	2 (33%)		
	Heartburn	3 (23%)	1 (17%)		
	Nausea	4 (31%)	2 (33%)		
	Vomiting	3 (23%)	2 (33%)		
Obstructive Symptoms	Blockage Sensation	Liquids	4 (31%)	2 (33%)	6 (32%)
		Liquids	0 (0%)	1 (17%)	0 (0%)
	Dysphagia	Thickened Liquids	0 (0%)	1 (17%)	1 (5%)
		Soft Foods	1 (7.5%)	1 (17%)	3 (16%)
		Dry Foods	3 (23%)	1 (17%)	7 (37%)
		Solids	3 (23%)	3 (50%)	8 (42%)
Change in Alimentary Habits	Need to drink	6 (46%)	4 (66%)	7 (37%)	
	Change in diet	6 (46%)	3 (50%)	11 (58%)	
	Last to finish meal	4 (31%)	3 (50%)	6 (32%)	
Pulmonary Symptoms (cough, pneumonia)	6 (46%)	3 (50%)	4 (21%)		



Dysmotility : Treatment

- Treat esophagitis +++
 - Prokinetic treatments acting on esophageal motor activity:
 - Cisapride
 - Bethanechol
 - Prucalopride
 - Prokinetics acting on gastric emptying:
 - Metoclopramide
 - Domperidone
 - Erythromycin
- Very disappointing

Miscellaneous and bizarre findings

Eosinophilic esophagitis

Batres et al. JPGN 2002
Gorter et al. JPS 2012



“Mucosal Bridge”

Boy, 10 years, EA type C, dysphagia



“Mucosal Bridge”

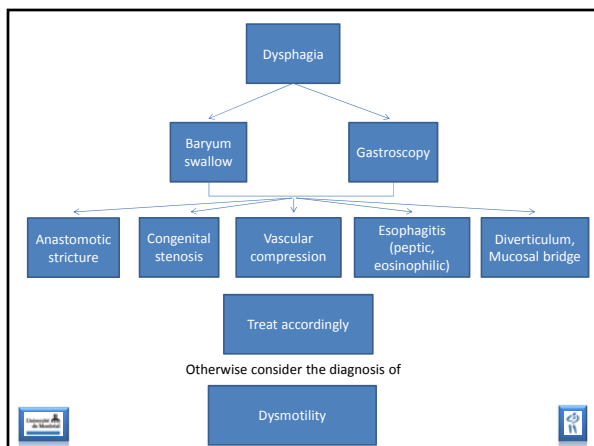


"Mucosal Bridge"



Diverticulum





Summary

- Dysphagia after EA repair is frequent
- Anastomotic stricture, esophagitis, congenital stenosis, vascular anomalies must be ruled out before incriminating dysmotility
- Dysmotility is universal in EA patients
- There is no correlation between dysmotility and symptoms
- Long term follow-up is mandatory including in adulthood to screen patients for Barrett's esophagus and esophageal cancer