

Tema 3. Atresia esofágica de long gap

Atresia de esófago

Planificar momento y tipo de cirugía



Prematuridad/Bajo peso
Dificultad ventilatoria
Cardiopatía
Malformaciones asociadas
Long Gap

N

Paciente estándar

Tratamiento en un tiempo

Decisión periodo neonatal

Neonatólogo
Cirujano
Anestesista

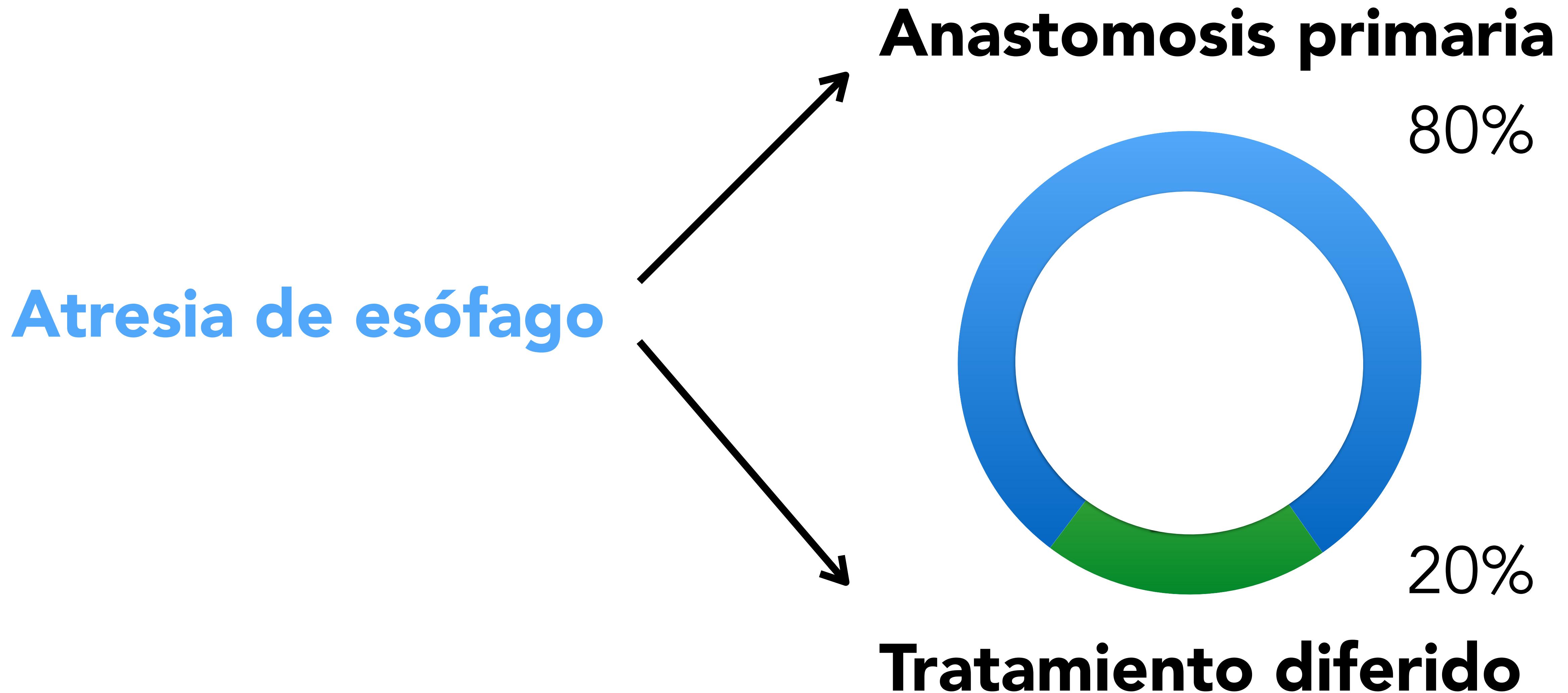


S

Paciente especial

Tratamiento diferido

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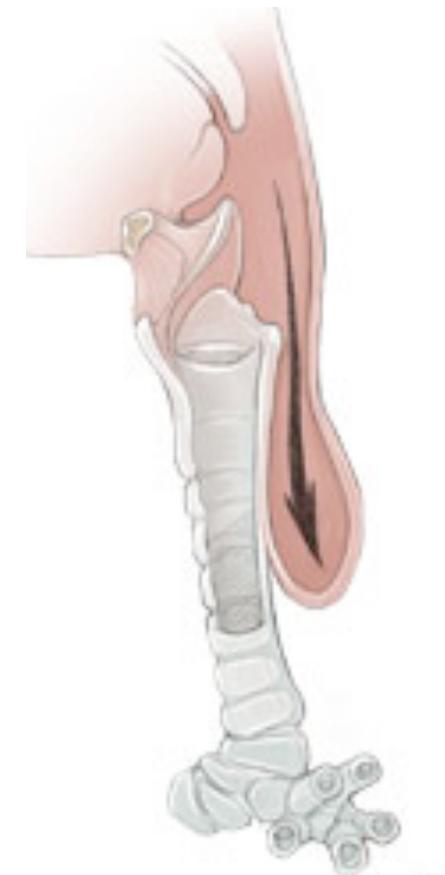


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¿Cual tipo de atresia puede ser long gap?

Tipo A



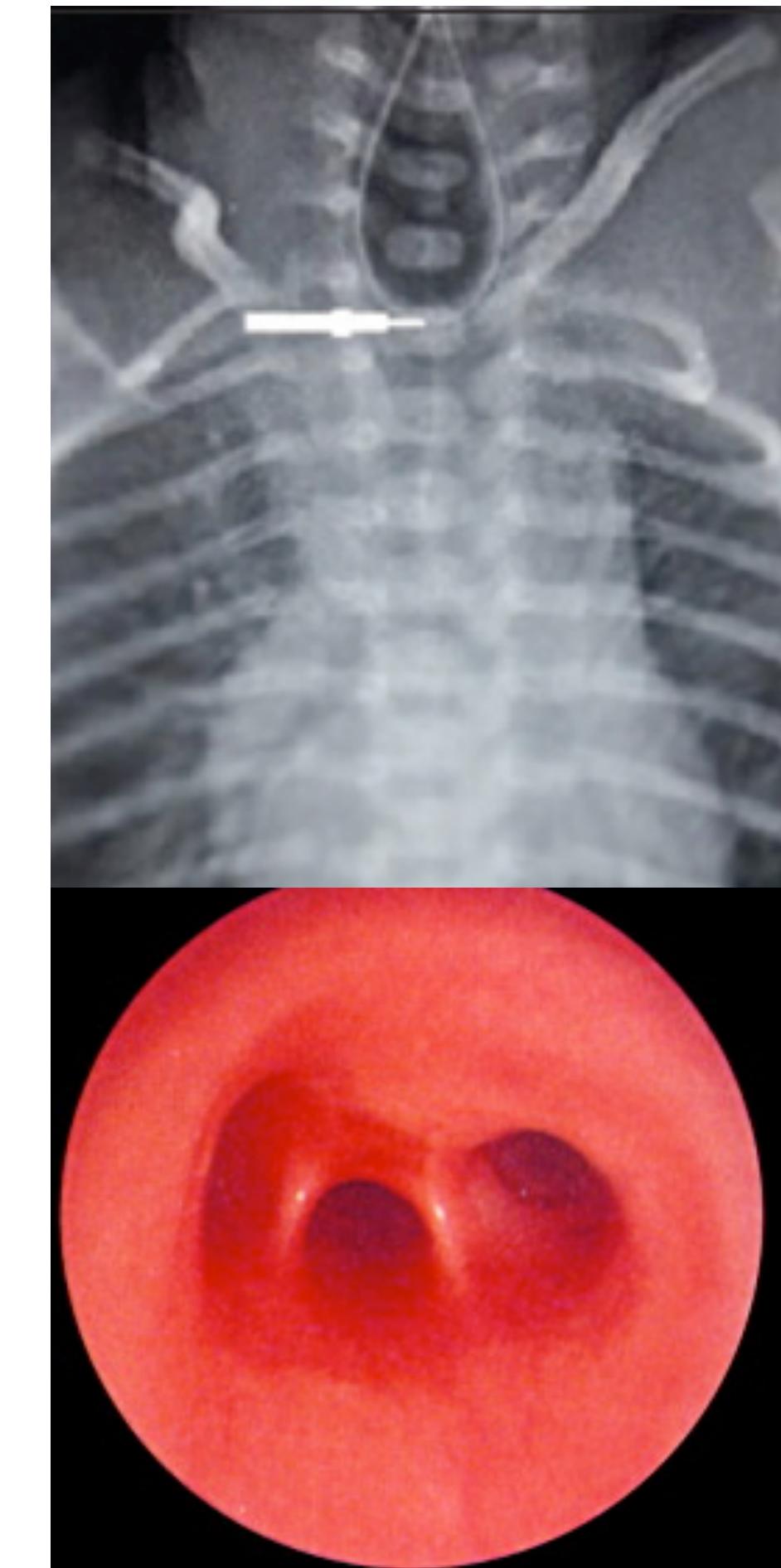
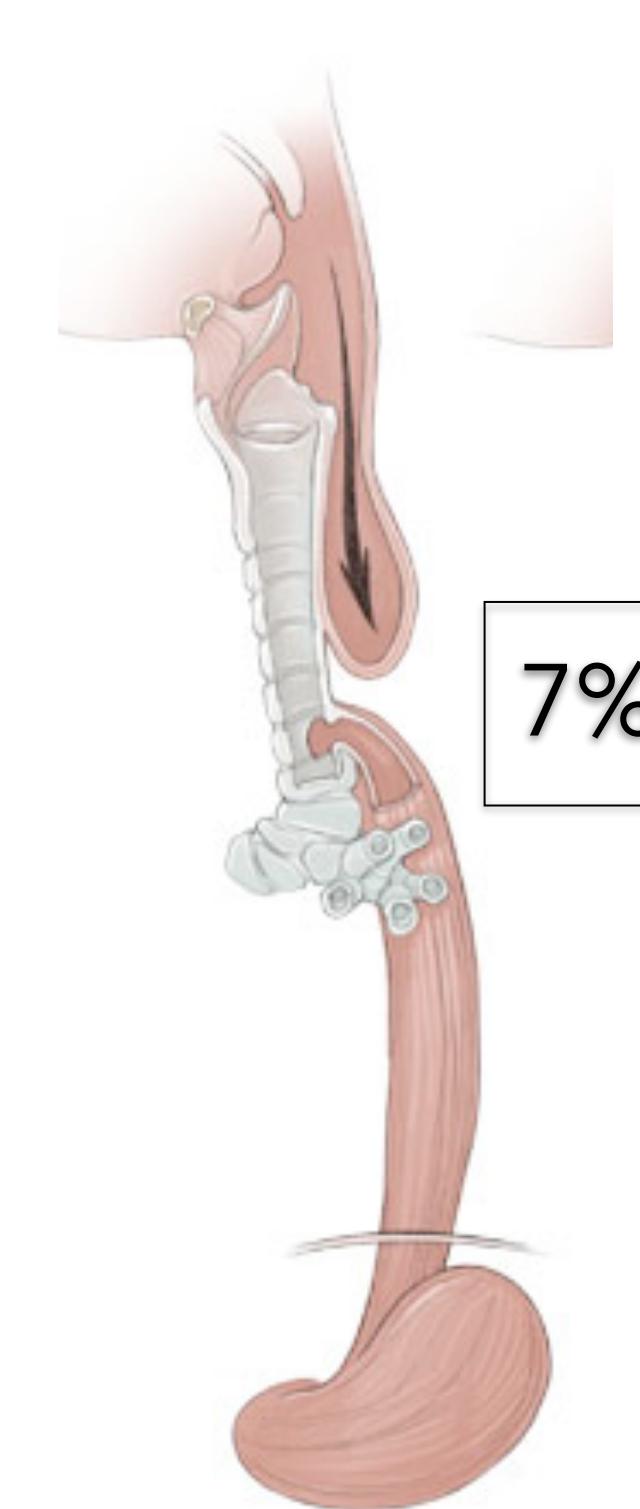
10%

Tipo B



2%

Tipo C



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¿Que se considera un long gap?

Authors – year	Long gap definition
Foker <i>et al.</i> – 1997 ¹¹	>2.5 cm
Hirschl <i>et al.</i> – 2002 ¹⁵	>3 cm
Bagolan <i>et al.</i> – 2004 ¹⁶	<u>>3</u> cm or vertebral bodies
Spitz – 2006 ¹⁷	>6 vertebral bodies
Sri Paran <i>et al.</i> – 2007 ¹⁸	Pure esophageal atresia
Hadidi <i>et al.</i> – 2007 ¹⁹	>4–5 vertebral bodies
Upadhyaya <i>et al.</i> – 2007 ¹⁴	>2.1 cm (ultra long gap if >3.5 cm)
Al-Shanafey and Harvey – 2008 ²⁰	... ‘when primary anastomosis was not possible’ ...

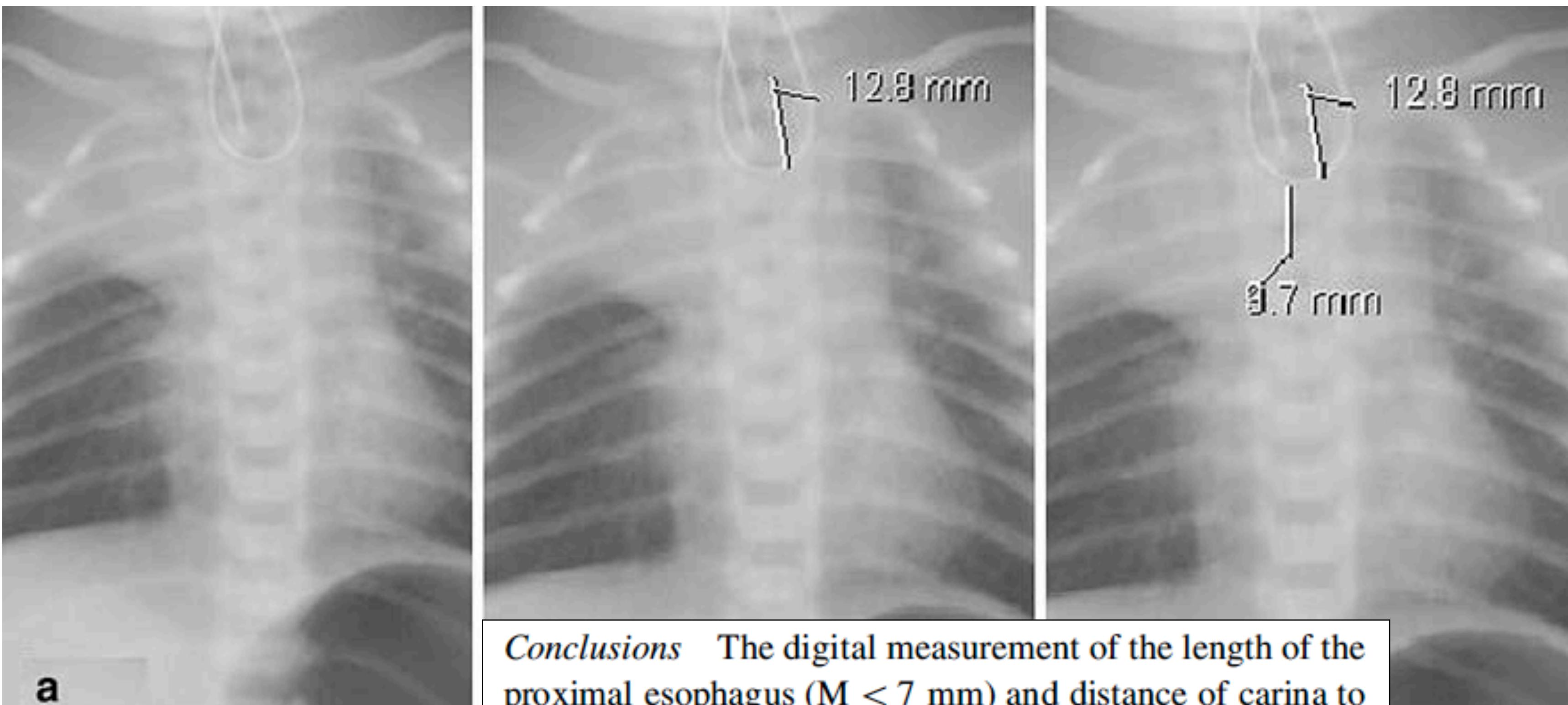
Concepto individual

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Complicaciones del long gap

A Novel Technique for Risk Calculation of Anastomotic Leakage after Thoracoscopic Repair for Esophageal Atresia with Distal Fistula

David C. van der Zee · Daisy Vieirra-Travassos ·
Justin R. de Jong · Stefaan H. A. J. Tytgat
World J Surg (2008) 32:1396–1399



Conclusions The digital measurement of the length of the proximal esophagus ($M < 7$ mm) and distance of carina to proximal esophagus ($M > 13.5$ mm) with the use of PACS gives a good risk calculation for postoperative leakage.

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Consecuencias de un long gap

Prognosis of congenital tracheoesophageal fistula with esophageal atresia on the basis of gap length

Vijay D. Upadhyaya · A. N. Gangopadhyaya ·
D. K. Gupta · S. P. Sharma · Vijayendra Kumar ·
Anand Pandey · Ashish D. Upadhyaya

Pediatr Surg Int (2007) 23:767–771

Gap length	No. of cases	Anastomotic leak	Stricture
Ultralong (>3.5 cm)	5	4 (80%)	1 (100%)
Long gap (>2 to <3.5 cm)	8	4 (50%)	3 (75%)
Intermediate gap (>1 to <2 cm)	18	5 (28%)	3 (22.5%)
Short gap (<1 cm)	19	2 (10.5%)	3 (19%)
Total (<i>n</i> = 50)	50	15 (30%)	10 (29%)

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¿Que conlleva un long gap?



- Reparación esofágica diferida
- Tratamiento prolongado
- Precisará de más de una intervención
- Mayor morbimortalidad
- No existe un tratamiento unificado**

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Incidencia de AE long gap

Prevalencia AE

1,8-2,4/10.000

Natalidad España 2016

(INE) 400.000 niv

72-96
casos AE

47 unidades públicas CIRUGÍA PEDIÁTRICA
1,8 - 2,4 casos/año

LONG GAP 10%: 7-9 casos por año

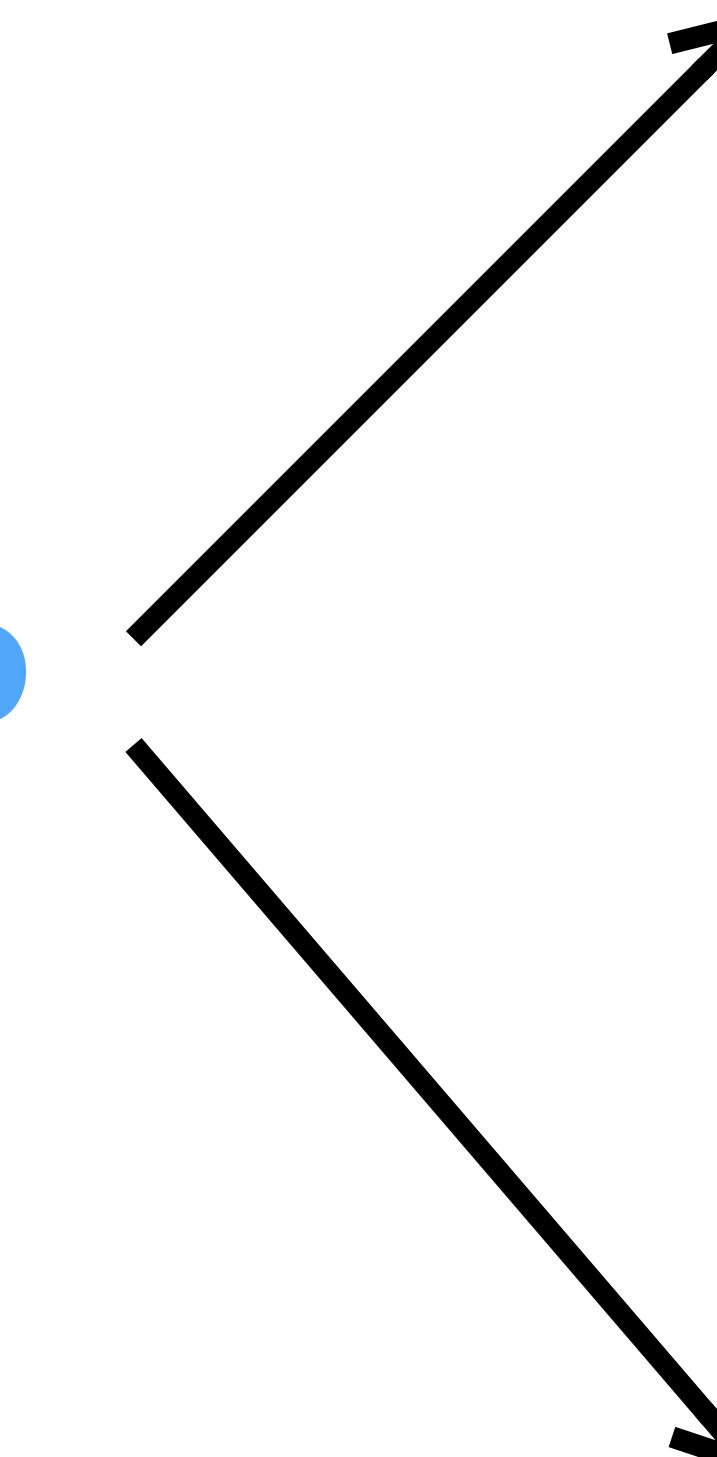


un caso cada cinco años por cada servicio

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Preservación esofágica

Tratamiento atresia long gap



Sustitución esofágica

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Técnicas en atresias long gap

■ Preservación esofágica

- Elongación esofágica pasiva (Puri)
- Elongación esofágica activa
 - Externa (Kimura)
 - Interna externa (Fokker)
 - Interna (Patowsky)

■ Sustitución esofágica

- Estómago
- Intestino delgado
- Cíolon

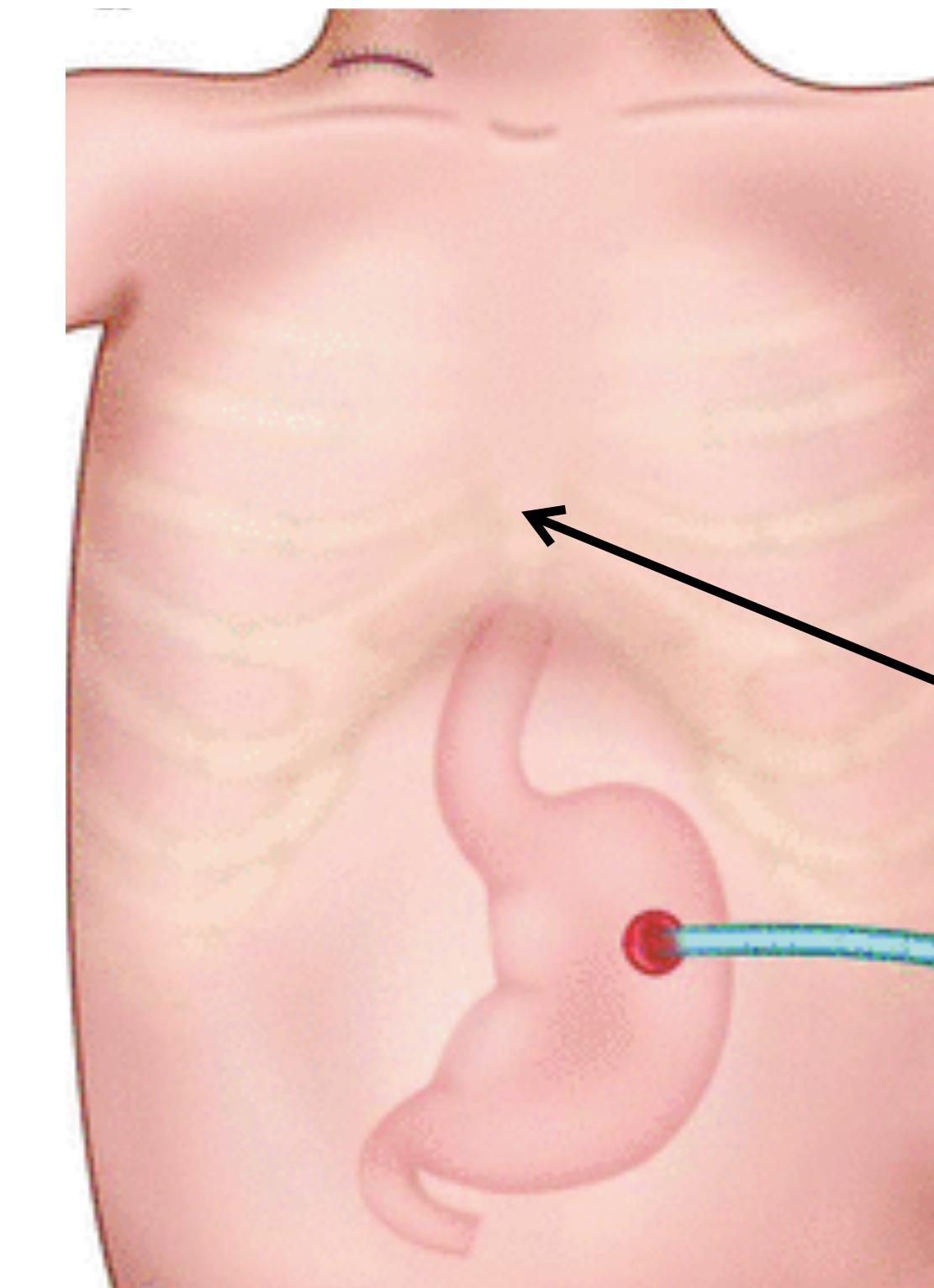
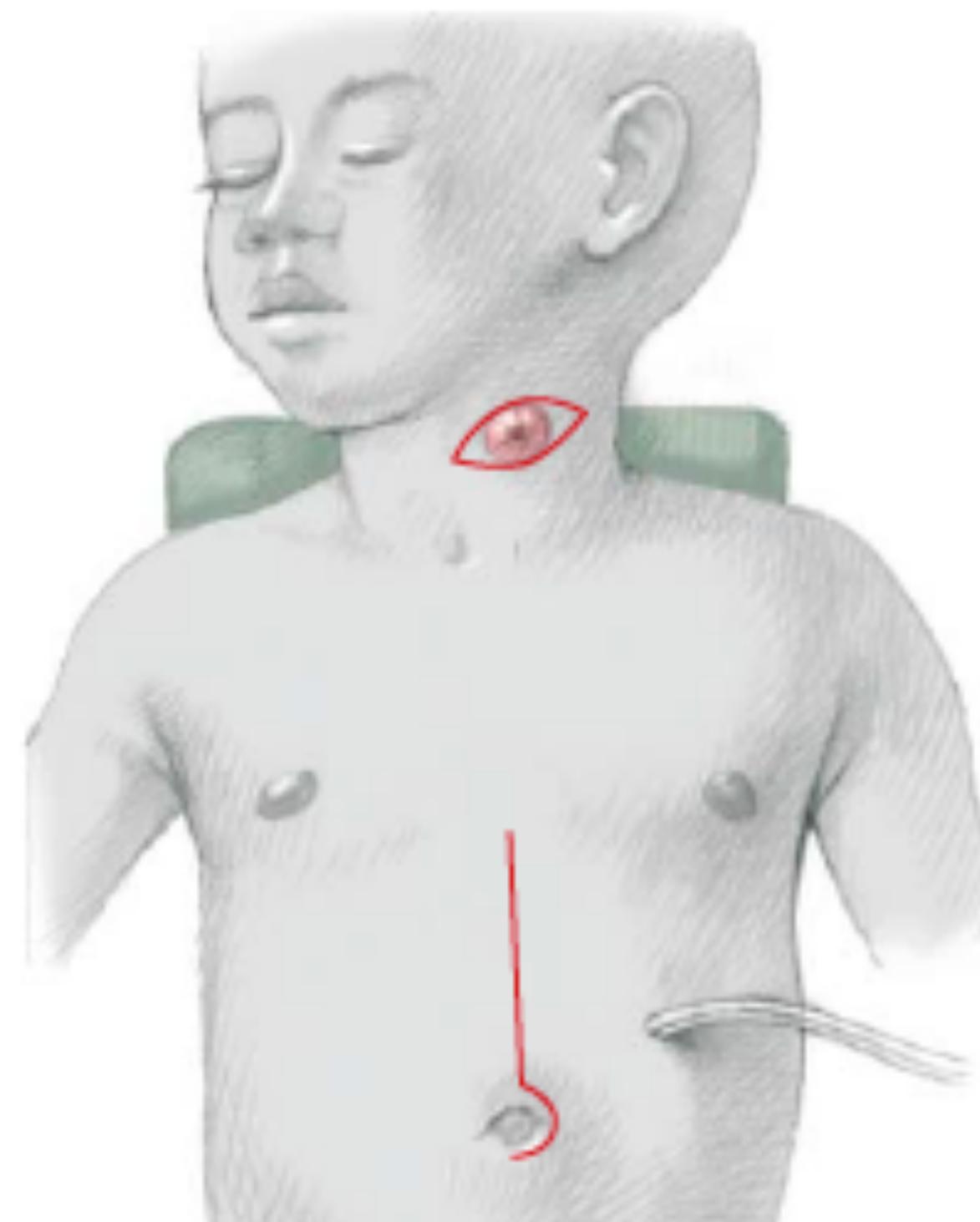
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Sustitución esofágica

(lo primero que se hizo, lo último que se debe hacer,

1.- Al nacimiento



2.- Al año

Cólon
Estómago
Intestino delgado

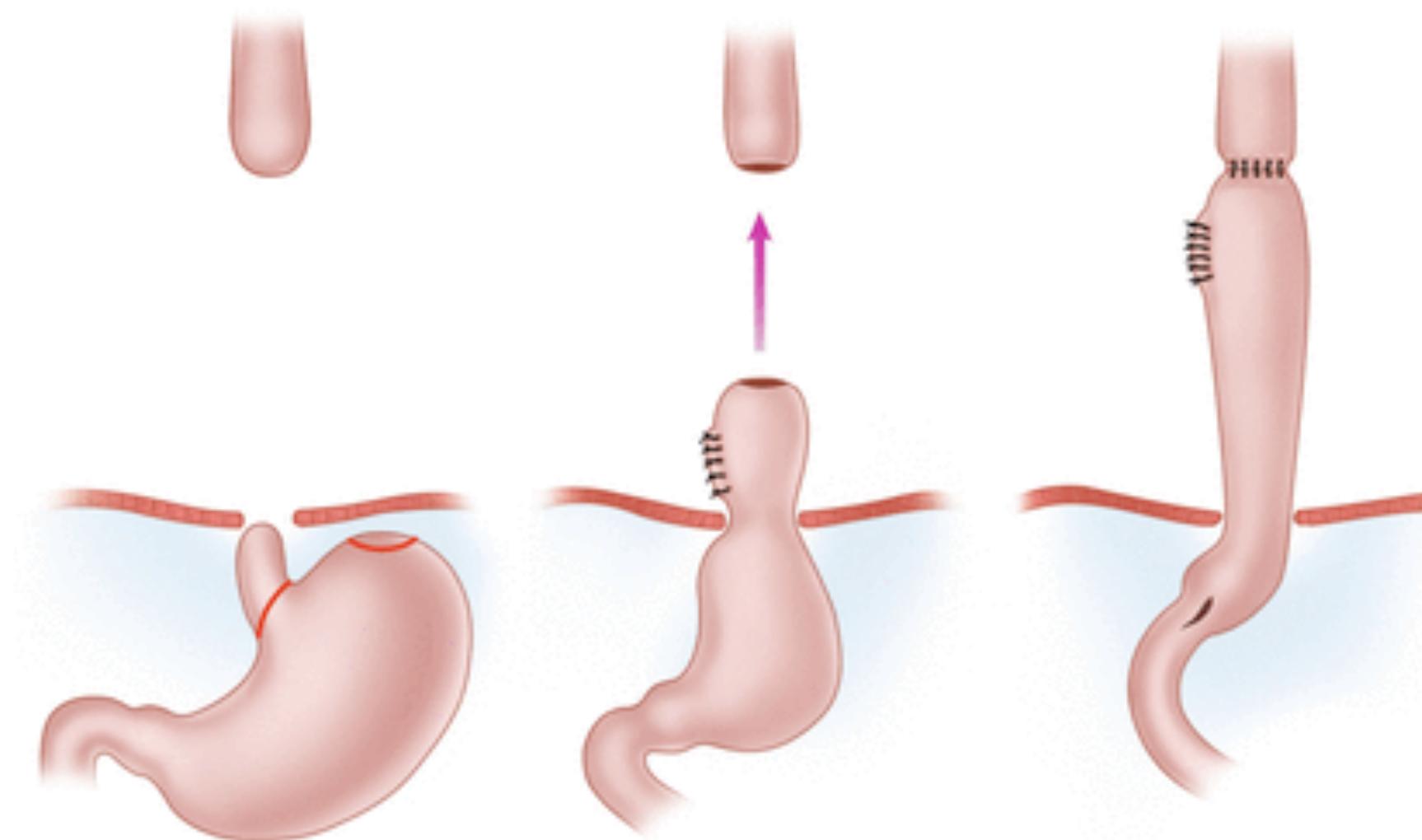
Esofagostomía + Gastrostomía

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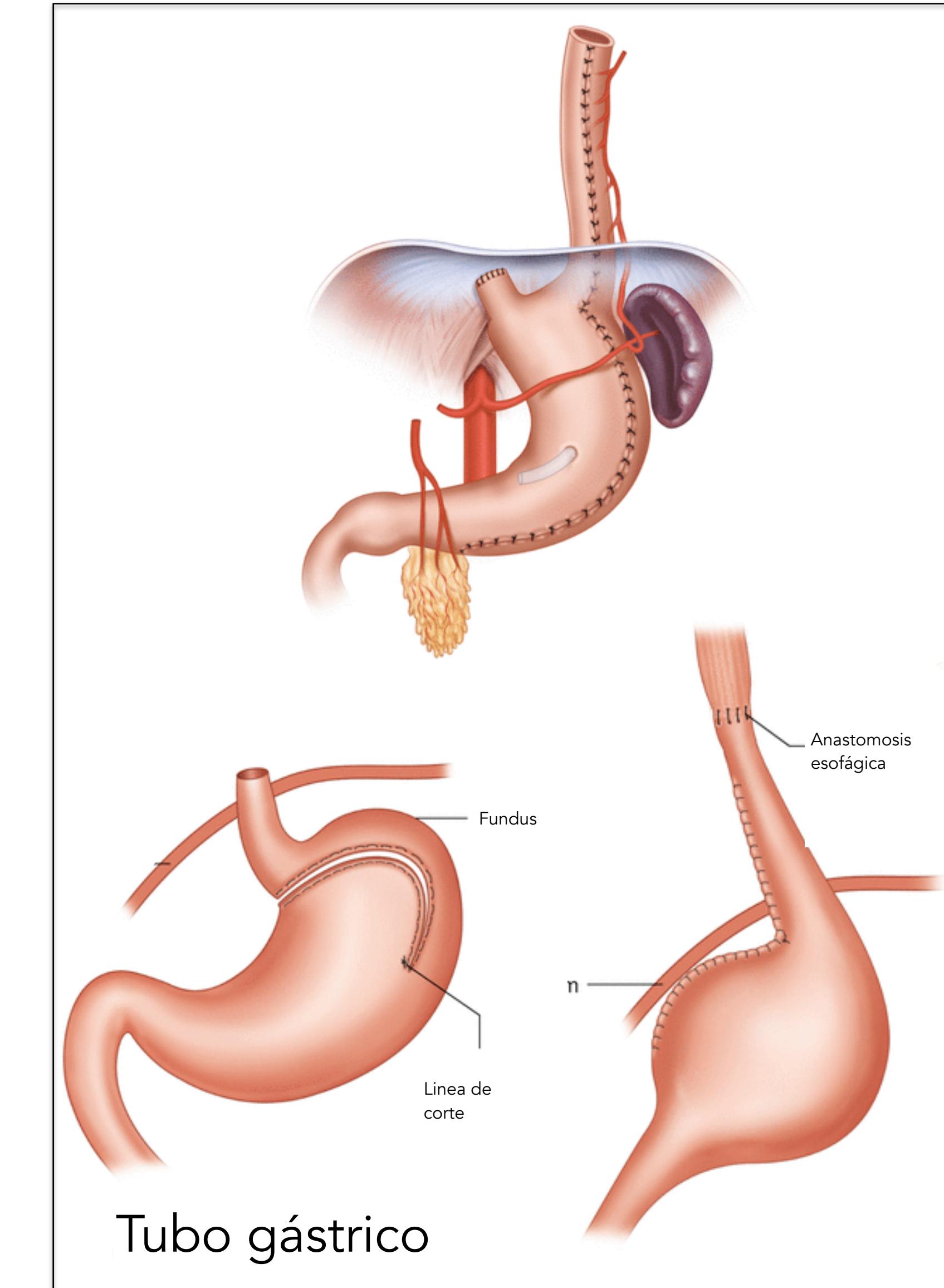
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Sustitución esofágica

Estómago



Ascenso gástrico



Tubo gástrico

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Sustitución esofágica

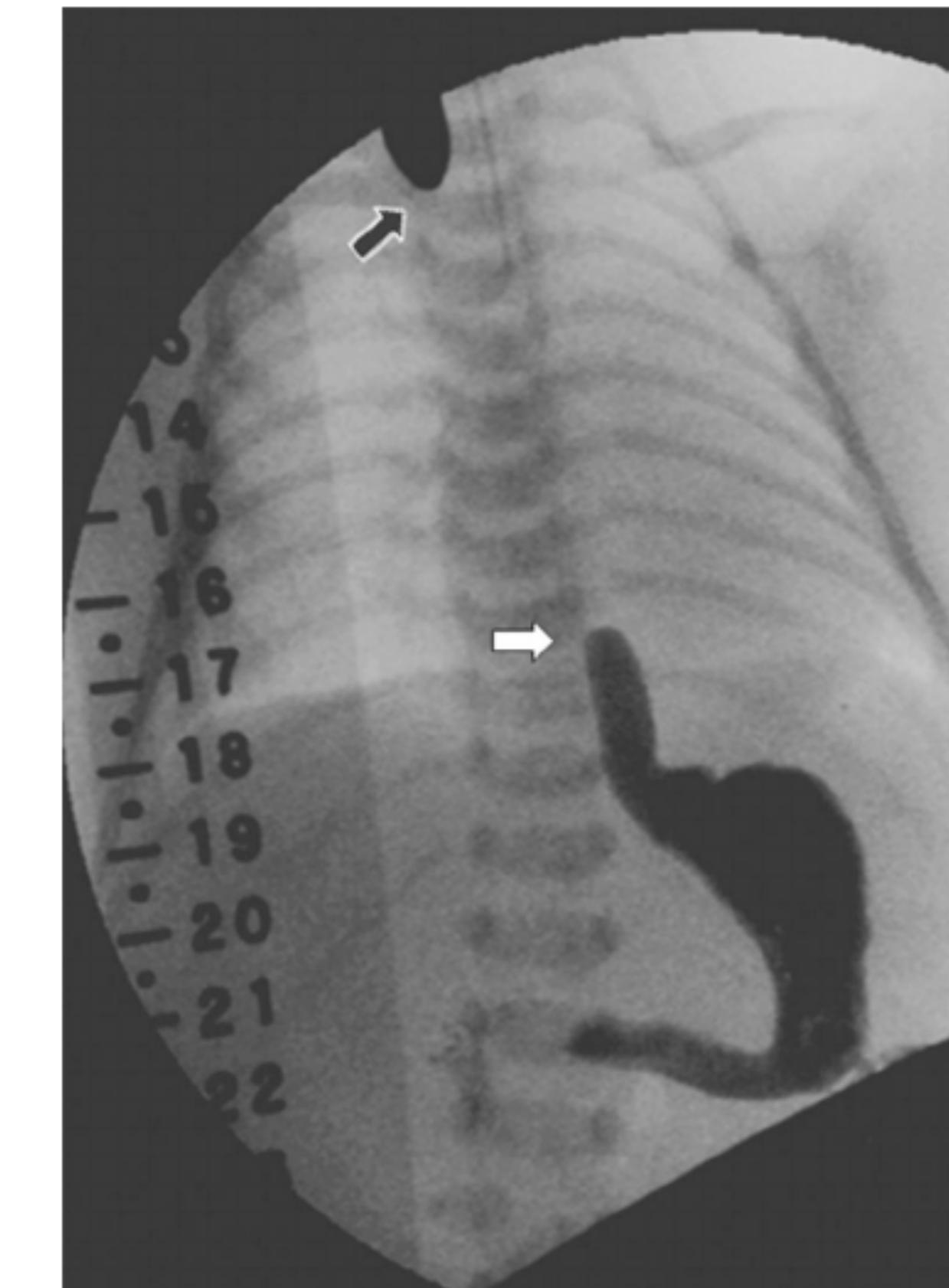
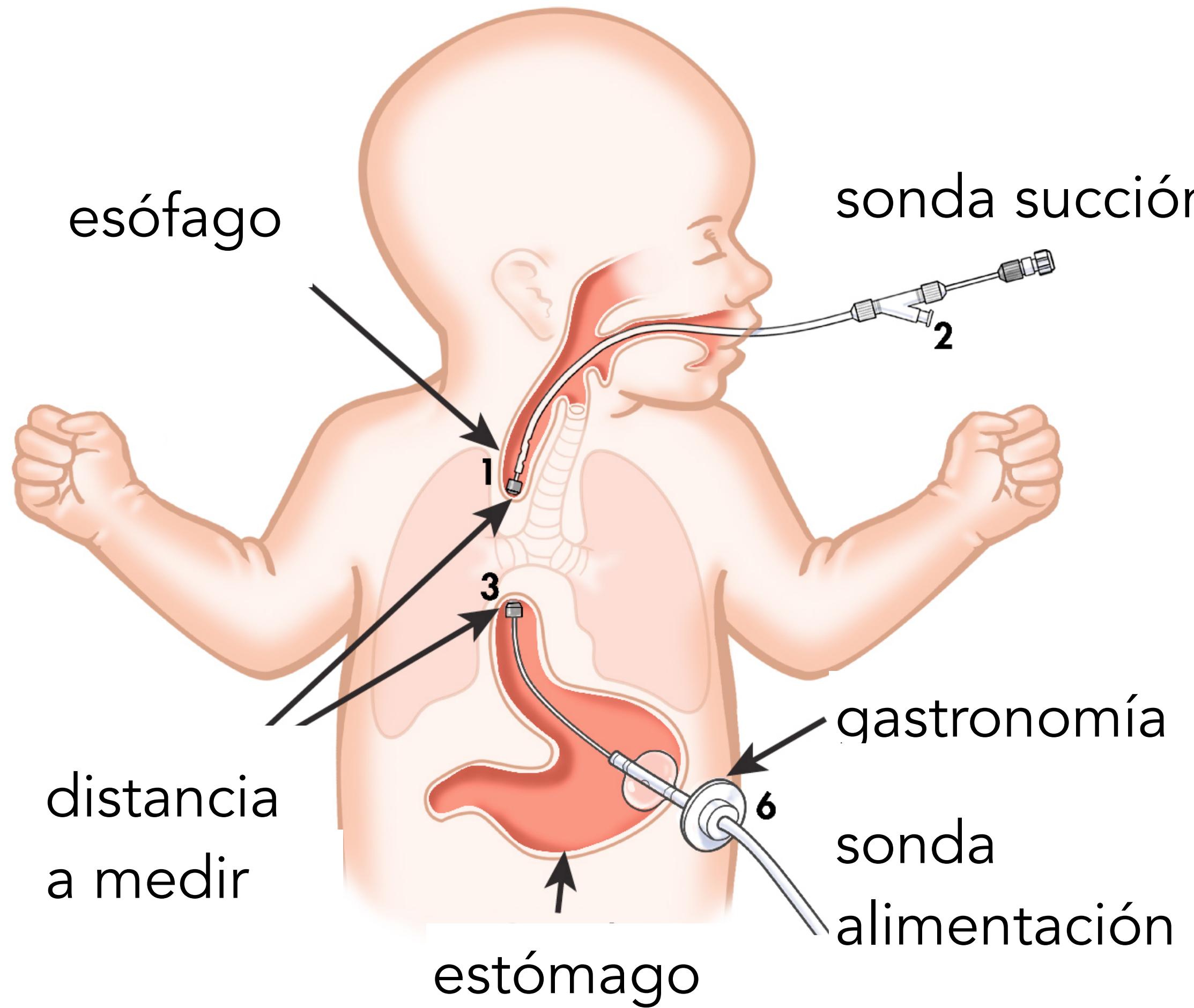
Cólon



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Elongación esofágica pasiva (Puri)



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Elongacion esofagica pasiva (Puri)



3 semanas

Crecimiento
espontáneo
del esófago

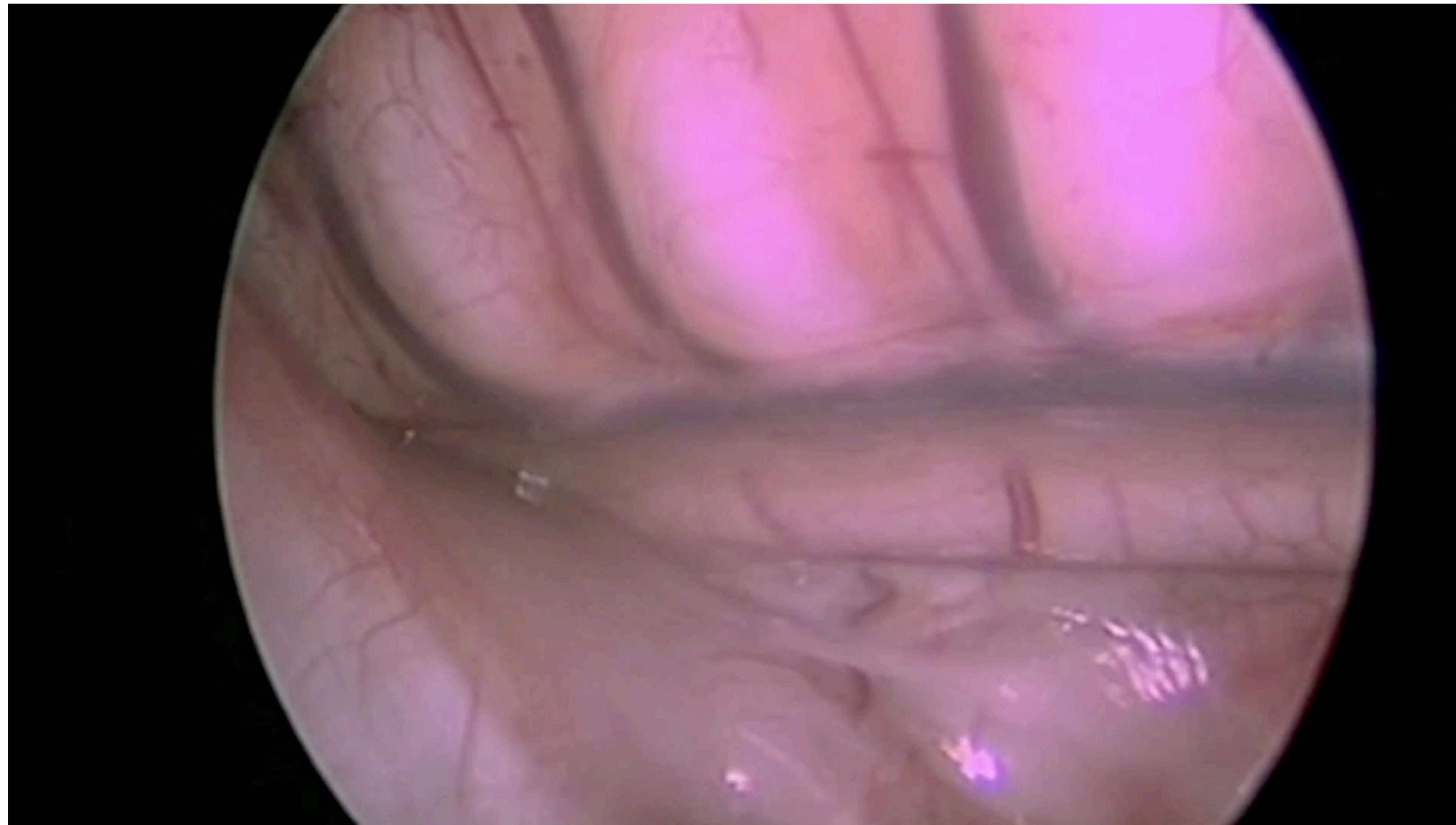


Mediciones
seriadas → 18 semanas

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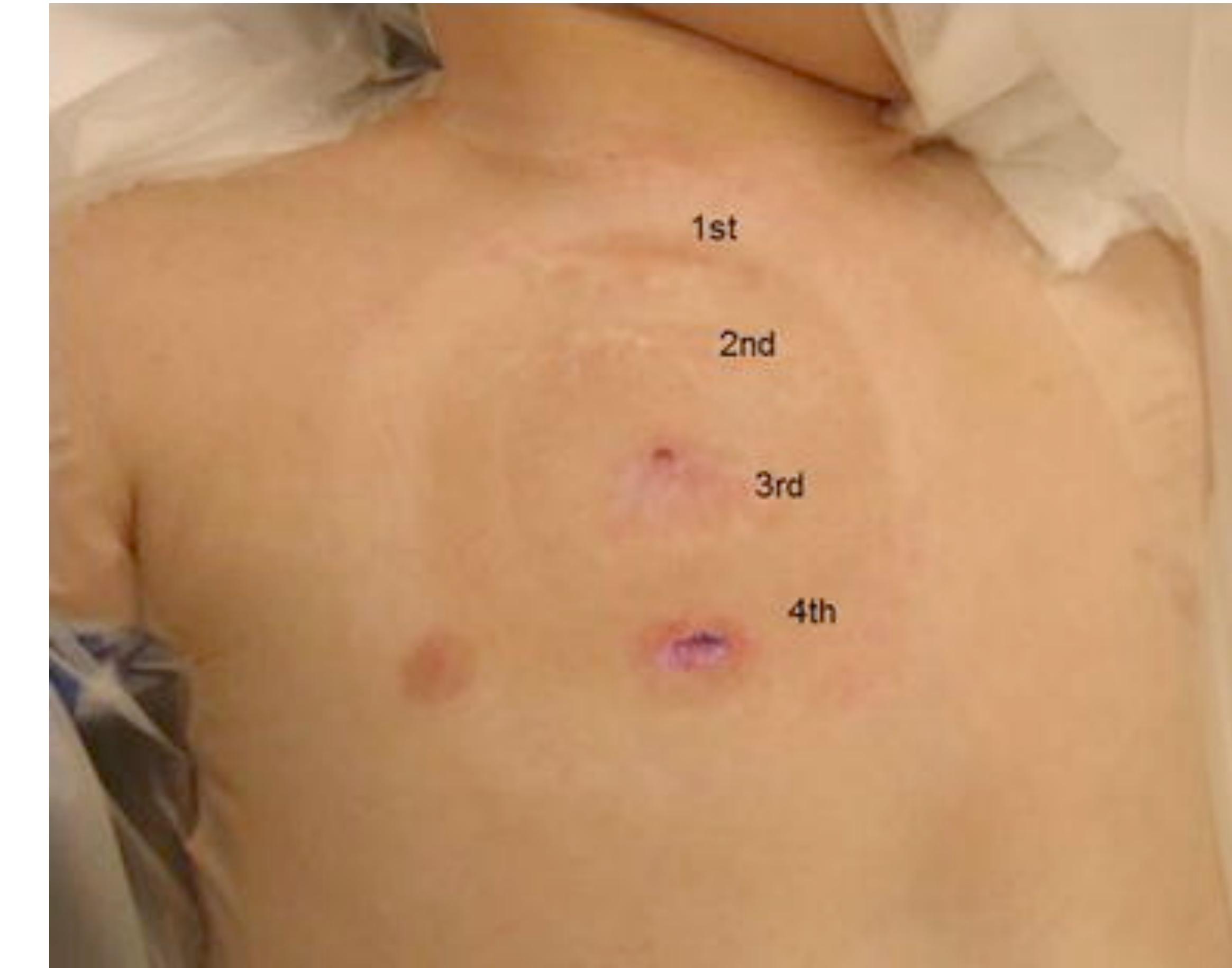
Elongación esofágica pasiva (Puri)



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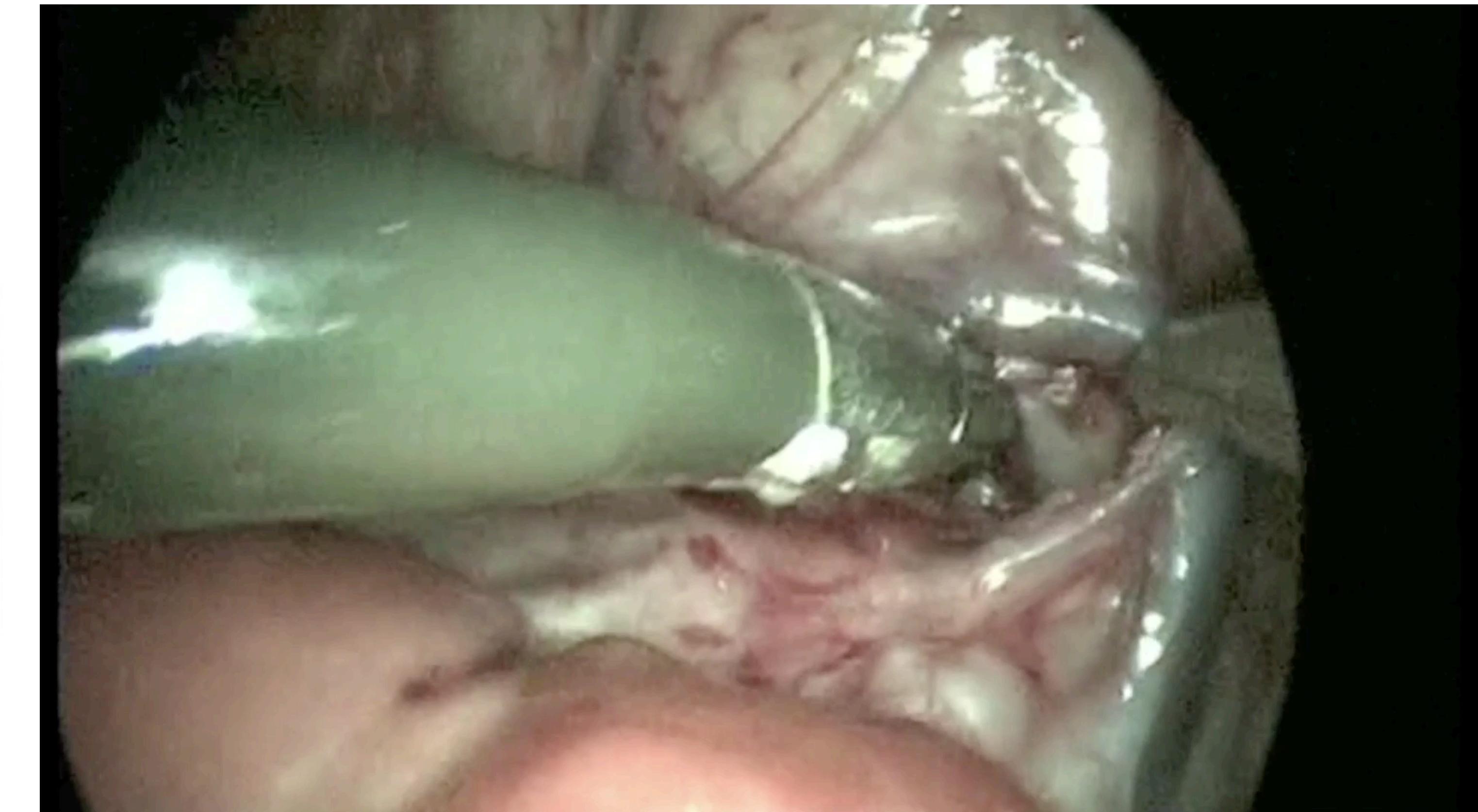
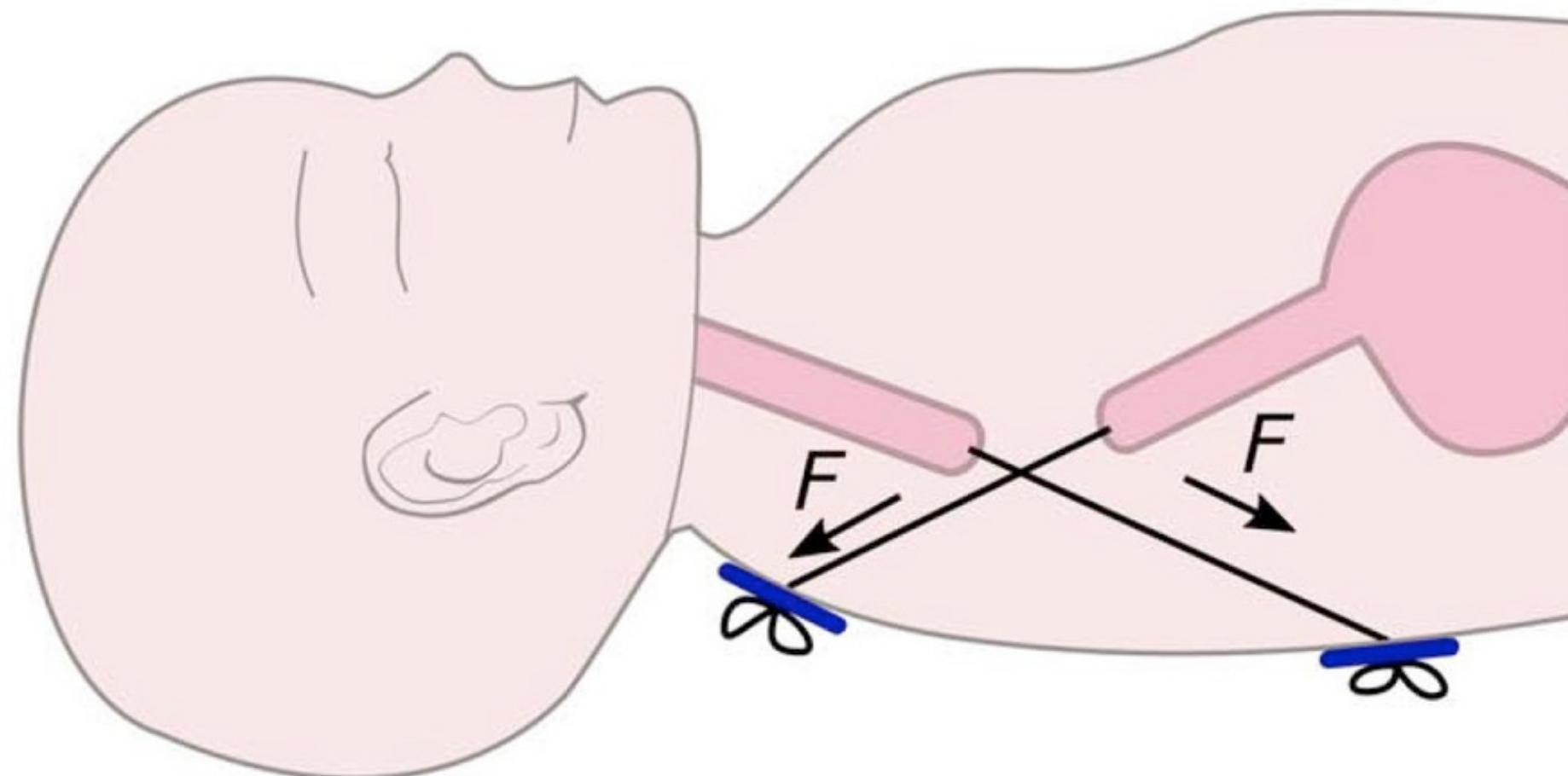
Elongación esofágica activa externa (Kimura)



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Elongación esofágica activa interno-externa (Fokker)

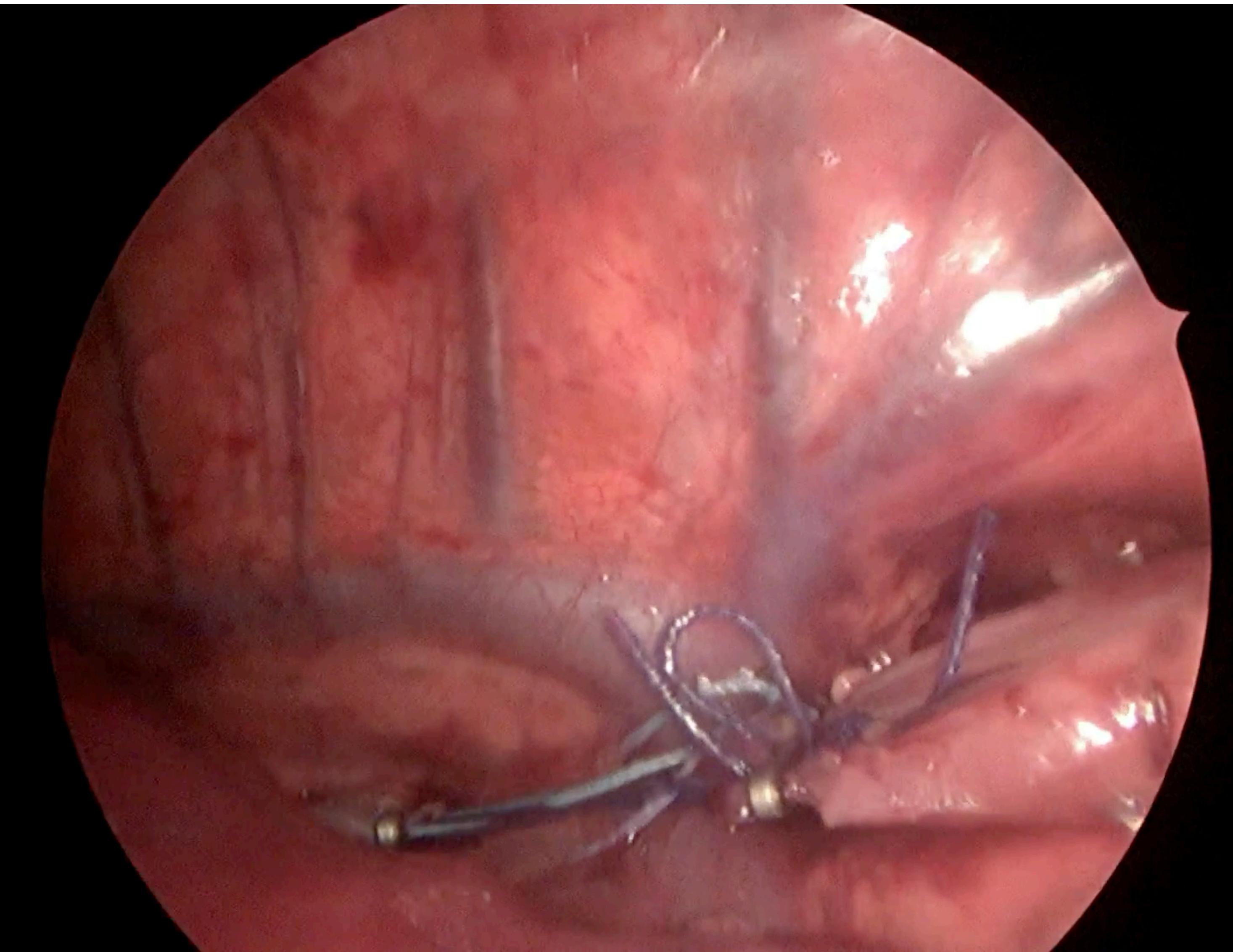
Fokker (toracotomía)



Fokker (toracoscopia)

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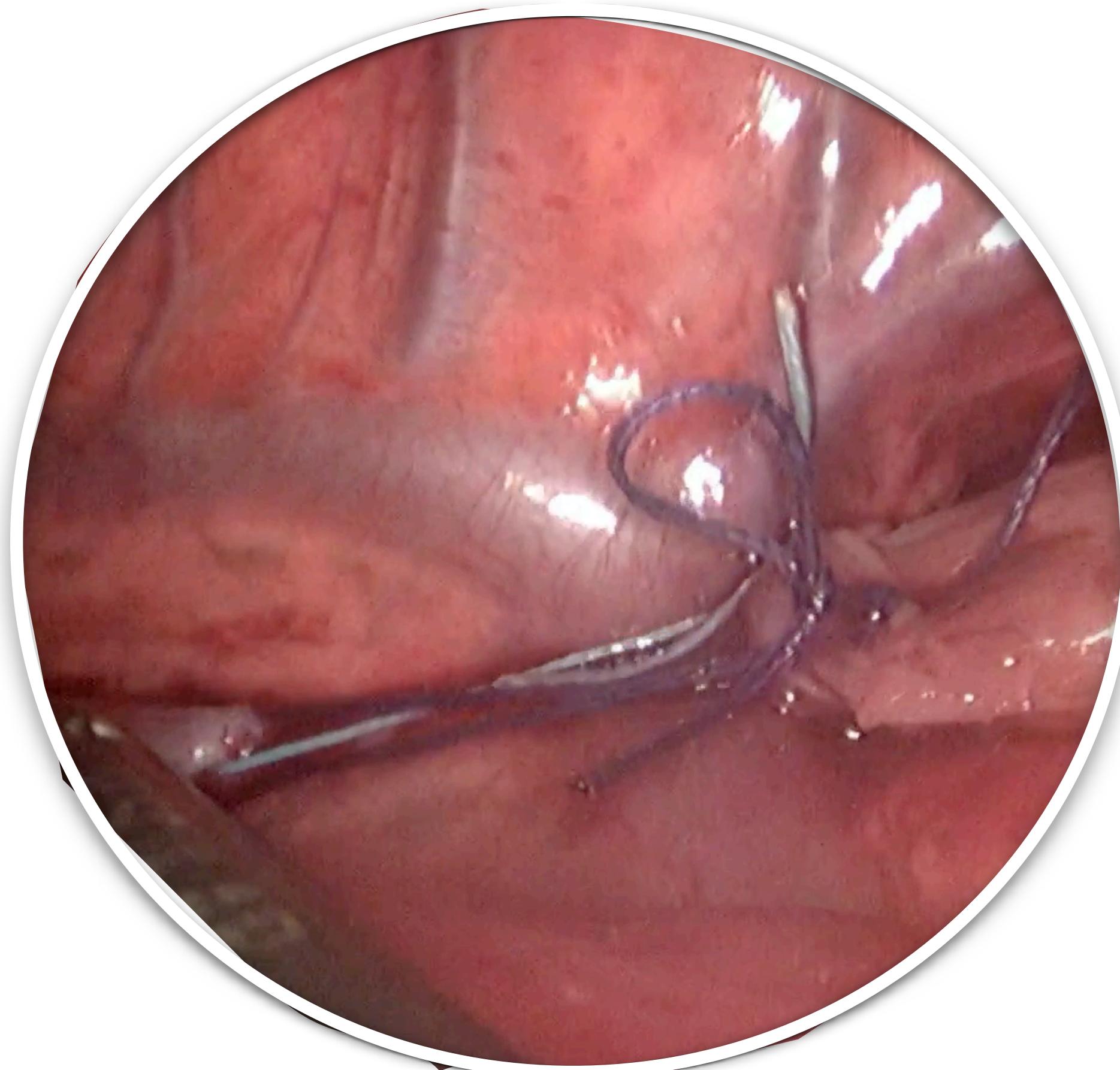
Elongación esofágica activa interna (Patowsky)



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Elongación esofágica activa interna (Patowsky)

- No se realiza gastrostomía (RGE)
- 2-3 intervenciones, cada 4 días
- Sedación prolongada



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	Técnica neonatal	Problema	Complicaciones	Resultados positivos
Puri Anastomosis diferida	Gastrostomia sin esofagostomia	Ingreso prolongado 3m	Aspiración Infecciones	70 %
Foker Elongación interna	Gastrostomia sin esofagostomia	Tracción no controlada	Rotura esófago	60 %
Patowsky Elongación interna	No gastrostomia No esofagostomia	Técnicamente compleja Varias cirugías	Ascenso gástrico	80 %

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J Pediatr Surg. 2019 Apr;54(4):675-687.

[Management of long gap esophageal atresia: A systematic review and evidence-based guidelines from the APSA Outcomes and Evidence Based Practice Committee](#)

Robert Baird 1 , Dave R Lal 2 , Robert L Ricca 3 , Karen A Diefenbach 4 , Cynthia D Downard 5 , Julia Shelton 6 , Stig Sømme 7 , Julia Grabowski 8 , Tolulope A Oyetunji 9 , Regan F Williams 10 , Tim Jancelewicz 10 , Roshni Dasgupta 11 , L Grier Arthur 12 , Akemi L Kawaguchi 13 , Yigit S Guner 14 , Ankush Gosain 15 , Robert L Gates 16 , Juan E Sola 17 , Lorraine I Kelley-Quon 18 , Shawn D St Peter 19 , Adam Goldin 20

Results: More than 3000 publications were reviewed, with 178 influencing final recommendations. In total, 18 recommendations are provided, primarily based on level 4-5 evidence. These recommendations provide detailed descriptions of the definition of LGEA, treatment techniques, outcomes and future directions of research.

Conclusions: Evidence supporting best practices for LGEA is currently low quality. This review provides best recommendations based on a critical evaluation of the available literature. Based on the lack of strong evidence, prospective and comparative research is clearly needed.

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Practice Guideline. Eur J Pediatr Surg. 2021 Jun;31(3):214-225.

ERNICA Consensus Conference on the Management of Patients with Long-Gap Esophageal Atresia: Perioperative, Surgical, and Long-Term Management.

Carmen Dingemann 1 , Simon Eaton 2 , Gunnar Aksnes 3 , Pietro Bagolan 4 , Kate M Cross 5 , Paolo De Coppi 2 5 , JoAnne Fruithof 6 , Piergiorgio Gamba 7 , Imeke Goldschmidt 8 , Frederic Gottrand 9 , Sabine Pirr 10 , Lars Rasmussen 11 , Rony Sfeir 12 , Graham Slater 13 , Janne Suominen 14 , Jan F Svensson 15 , Joergen M Thorup 16 , Stefaan H A J Tytgat 17 , David C van der Zee 17 , Lucas Wessel 18 , Anke Widenmann-Großig 19 , René Wijnen 20 , Wilhelm Zetterquist 21 , Benno M Ure 1

Results: Ninety-seven items were generated. Complete consensus (100%) was achieved on 56 items (58%), e.g., **avoidance of a cervical esophagostomy, promotion of sham feeding,** details of delayed anastomosis, **thoracoscopic pouch mobilization and placement of traction sutures as novel technique,** replacement techniques, and follow-up. Consensus $\geq 75\%$ was achieved on 90 items (93%), e.g., definition of long gap, routine pyloroplasty in gastric transposition, and avoidance of preoperative bougienage to enable delayed anastomosis. **Nineteen items (20%), e.g., methods of gap measurement were discussed controversially (range 1-9).**

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J. Surgical Research. 2020. (251) 47-52

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Outcomes for Correction of Long-Gap Esophageal Atresia: A 22-Year Experience.

Amanda R. Jensen, MD, MS,a,b,*Lucas A. McDuffie, MD,a,bEric M. Groh, MD,a,b and Frederick J. Rescorla, MDa,b,c

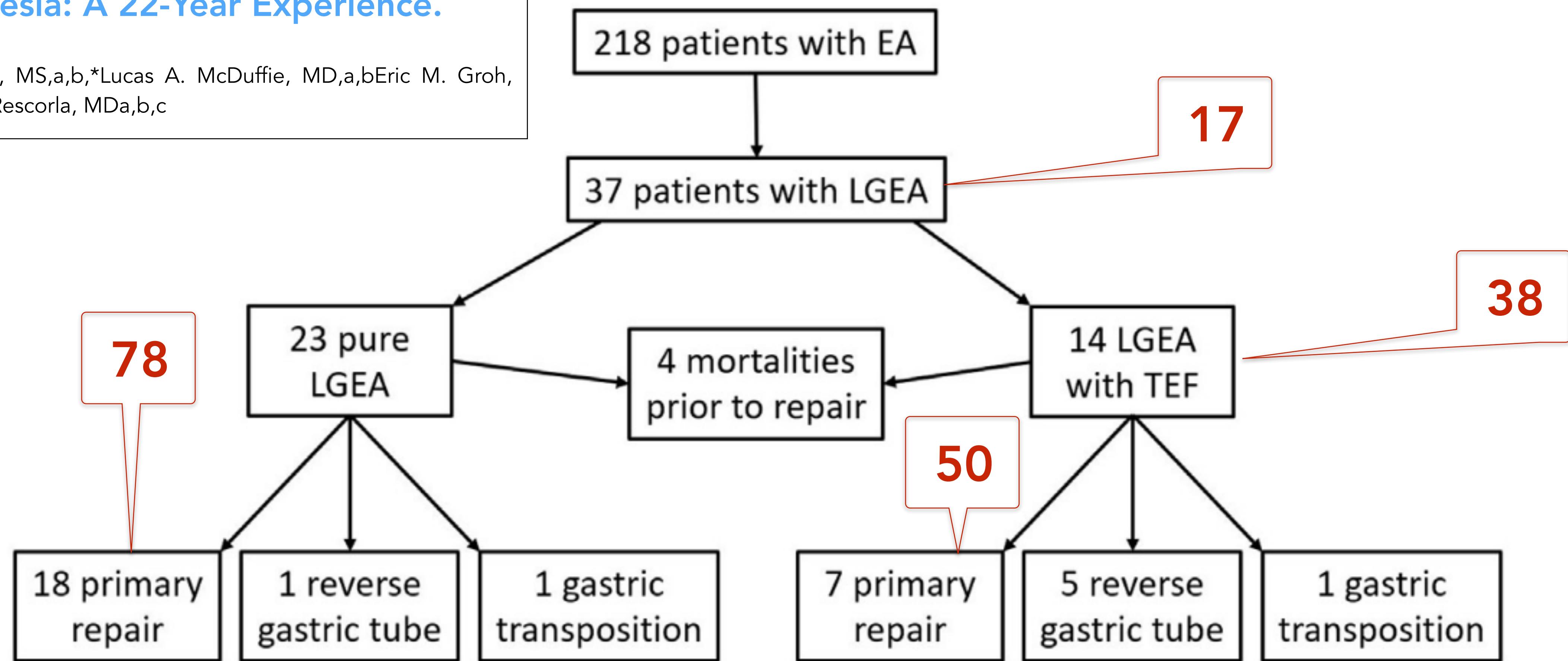


Fig. 2 – Flow diagram of patients with long gap esophageal atresia.

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Published: August 01, 2022 DOI: <https://doi.org/10.1016/j.jpedsurg.2022.07.023>

AgateBourga,*FrédéricGottrandb, BenoitParmentiera, JulieThomasa, AnneLehnc, ChristianPiolatd, ArnaudBonnaire, RonySfeirf, JulieLienardg, VéroniqueRousseauh, MyriamPouzaci, AgnèsLiardj, PhilippeBuissonk, AuroreHaffreinguel, LouisDavidm, SophieBranchereau, VéroniqueCarcauzono, NicolasKalfap,Marc-DavidLeclairq, HubertLardyr, Sabinelrtans, FrançoisVarlett,T homasGelasu, DianaPotopa, MarieAuger-Hunaulta

Outcome of long gap esophageal atresia at 6 years: A prospective case control cohort study

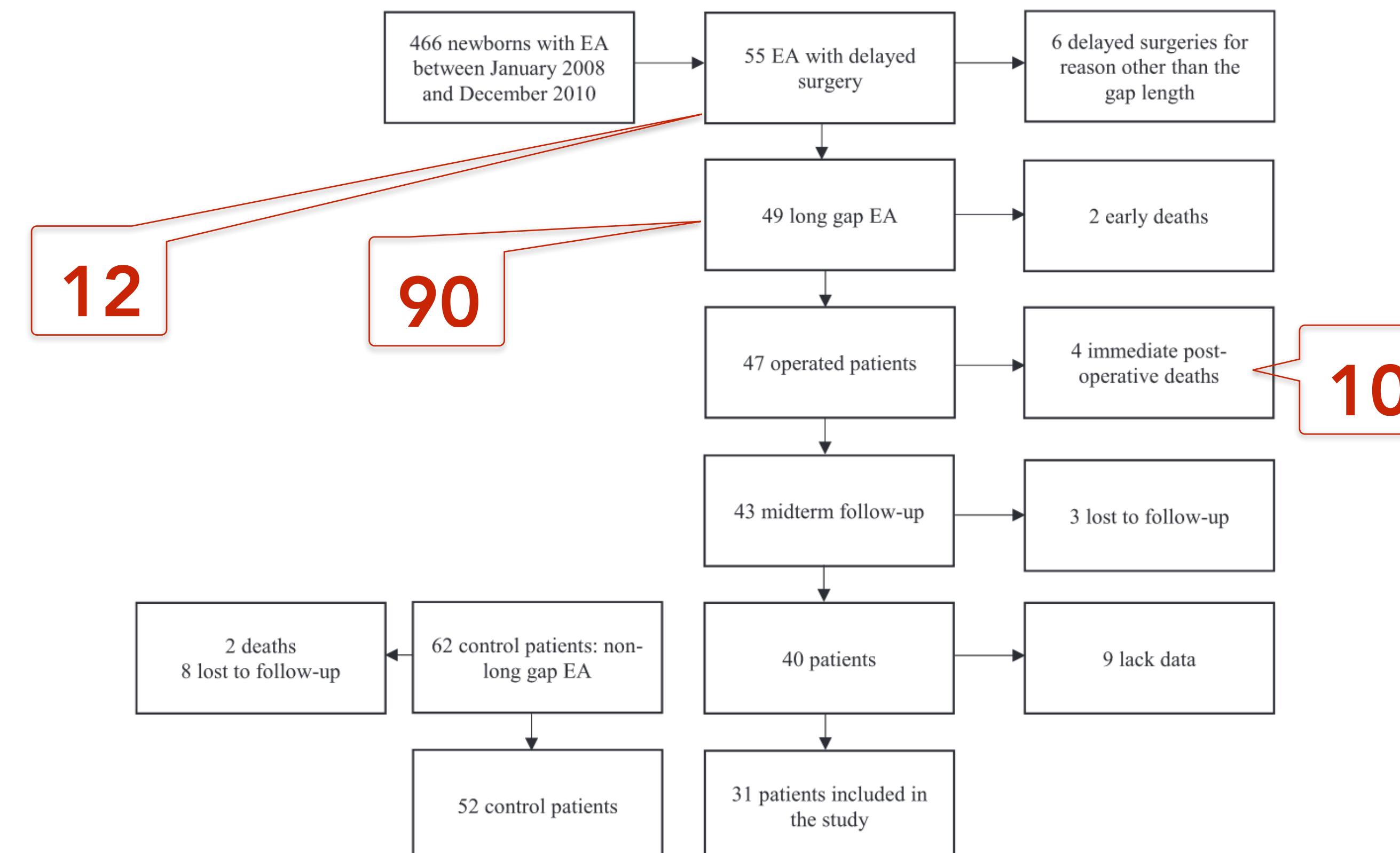


Fig. 1. flowchart of the study.

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Outcome of long gap esophageal atresia at 6 years: A prospective case control cohort study

Table 4

Characteristics and outcomes of “native esophagus conservation” and “esophagus replacement” in long gap EA group.

Characteristics	Native esophagus N = 33	Esophagus replacement N = 14	P-value
Median term in gestational age (range)	36 (26.8; 39.8)	37 (31; 41)	0.23
Median birth weight in grams (range)	2072 (1040; 3740)	2459 (550; 3025)	0.04 *
Associated anomalies (%)	15 (45)	9 (64)	0.34
Cardiac malformations (%)	7 (21)	4 (28)	0.46
VACTERL (%)	7(21)	4 (28)	0.46
Ladd classification (%)	type I 25 (76) type II 3 (9) type III 4 (12) type IV 1 (3)	type I 10 (71) type II 2 (14) type III 2 (14) type IV 0 (0)	0.46

13%

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Table 5

Outcomes at one year and 6 years of “esophagus replacement” and “native esophagus conservation” in long-gap EA group.

Characteristics	Esophagus conservation n = 32	Esophagus replacement n = 13	P-value
1 YEAR			
Weight (grams)	8128	8106	0.99
Readmission (%)	25 (78%)	13 (100%)	0.06
Number of readmission median (ranges)	2.5 (0; 6)	3 (1; 7)	0.96
Median readmission duration in days (extreme)	9 (0; 148)	13 (0; 114)	0.51
Total length of stay in days during the first year (extreme)	152 (86; 365)	160 (70; 365)	0.43
Anti-reflux surgery	17 (53%)	6 (46%)	0.67
GERD	25/32 (78%)	5/12 (42%)	0.009
Dysphagia	9/31 (29%)	3/12 (25%)	0.73
Exclusive oral feeding	16 (50%)	4 (31%)	0.25
Intercurrent respiratory event	9 (28%)	5 (38%)	0.74
Respiratory treatment	9/28 (32%)	6/13 (46%)	0.25
6 YEARS			
Lost to follow-up since age one year	10	4	
Weight (grams) mean	24.6 (SD)	24.4	0.9
FOIS (mean)	6	6	0.33
GERD	6	4	0.36
Undernutrition	16	8	0.34
Dysphagia	14	3	0.13
Asthma	8	2	0.46

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Protocolo de estandarización

- Rx. primera cirugía
 - Edad gestacional
 - Peso
 - Medida en cm/placa standard
 - Decisión según gap
-
- Resultados quirúrgicos
 - Dehiscencia
 - Refistulización
 - Estenosis

Sonda resistente
o tallo de Hegar con
presión moderda

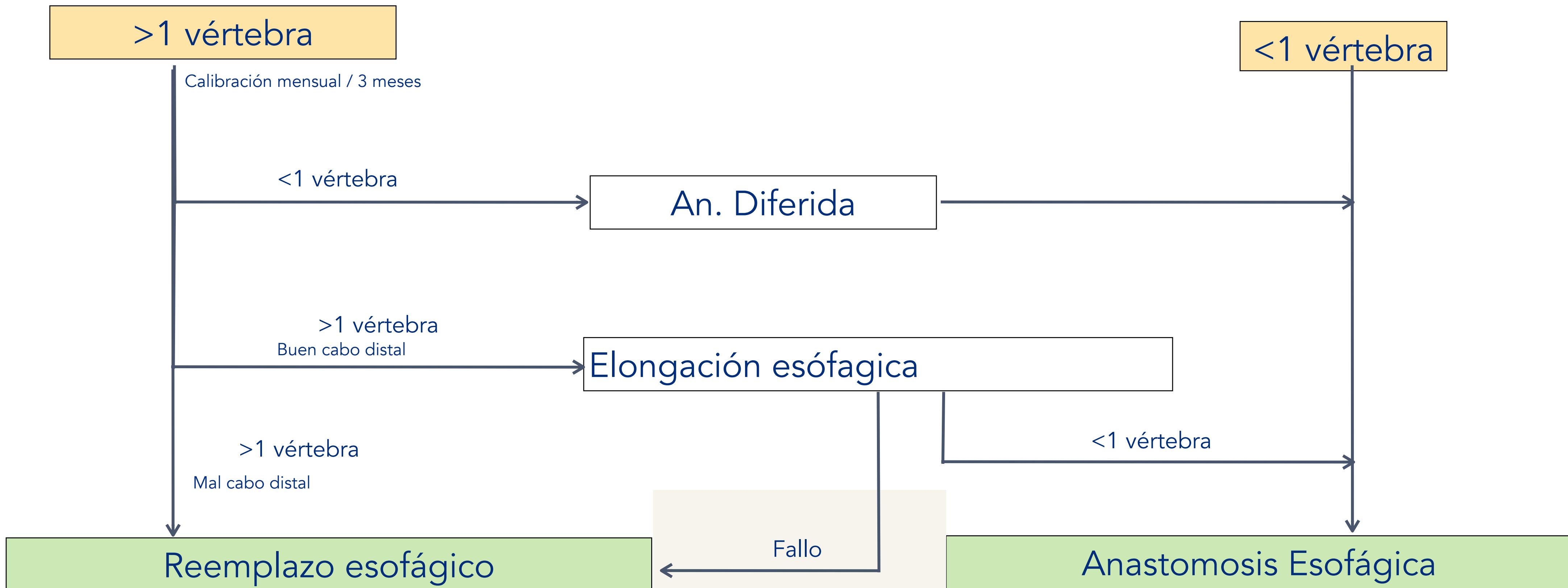
Broncoscopio
en la fístula



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Algoritmo. Atresia tipo A



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