

Ti ricordiamo che questo materiale
è di proprietà dell'Autore.
Come partecipante al
XXVIII CONGRESSO NAZIONALE
SIMRI questo materiale ti è fornito da
SIMRI per esclusivo uso personale
concesso dall'Autore



XXVIII CONGRESSO NAZIONALE SIMRI

Il respiro: scienza e terapia per la salute del bambino



Torino, 10-12 ottobre 2024

Follow-up nelle malattie respiratorie croniche:
malacia delle vie aeree

Dott. Pietro Salvati

UOC di Pneumologia Pediatrica ed Endoscopia Respiratoria
IRCCS Istituto Giannina Gaslini Genova



Conditions associated with TM–BM- TBM

TABLE 1 Summary of causes of tracheomalacia (TM), bronchomalacia (BM) and tracheobronchomalacia (TBM)

Primary or congenital			Secondary or acquired		
Congenital idiopathic Idiopathic TM/BM (may be genetic factors)			Cardiovascular anomalies associated with TM/BM		
Congenital abnormalities of the cartilage			Double aortic arch	Dilated cardiomyopathy	Pulmonary arterial sling
Dyschondroplasia/chondromalacia/achondroplasia	Ehlers–Danlos syndromes	Marfan syndrome	Right aortic arch	Aberrant right subclavian	Enlarged pulmonary veins
Left bronchial isomerism with normal atrial arrangement			Left atrial hypertrophy	Enlarged left atrium	Severe PAH
Congenital anomalies of the aerodigestive tract			Left to right shunting leading to enlarged pulmonary arteries	Tetralogy of Fallot with absent pulmonary valve syndrome	Abnormal take-off of the innominate artery
Oesophageal atresia (with or without laryngeal cleft)	TOF		Skeletal anomalies associated with TM/BM	Pectus excavatum	
Anomalies of respiratory tract development			Scoliosis		
Prematurity	Bronchopulmonary dysplasia		Infections and inflammatory processes associated with TM/TBM	Protracted bacterial bronchitis	Stevens–Johnson syndrome
Congenital syndromes associated with TM/TBM			Chronic suppurative lung disease, including cystic fibrosis, primary ciliary dyskinesia, other causes of bronchiectasis	Relapsing polychondritis	
Mucopolysaccharidosis (Hurler syndrome, Hunter syndrome)	CHARGE syndrome	VATER anomaly	Tracheobronchial injury associated with TM/TBM		
Trisomy 9	Trisomy 21	Cri du chat syndrome	Button battery ingestion injury	Delayed removal of inhaled foreign body	Trauma
Smith's syndrome	Opitz syndrome	Goldenhaar syndrome	Medical procedures and surgery associated with TM/TBM		
Cotello's syndrome	Neurofibromatosis	Allagille's syndrome	Prolonged intubation	Tracheostomy	TOF repair
Arthrogyposis	Atelosteogenesis type 1	18–22 translocation	Laryngotracheal reconstruction	Tracheoplasty	Heart transplant
Antley–Bixler syndrome; 11p13 deletion; 16p13.3 deletion; 22q11 deletion	Partial trisomy of long arms of chromosomes 11 and 22	Larsen syndrome and Larsen-like syndromes	Fetal balloon insertion for congenital diaphragmatic hernia		
Pfeiffer syndrome	Blackfan–Diamond anaemia	Williams–Campbell syndrome	Tumours and cysts associated with TM/TBM		
Kniest dysplasia	Diastrophic dysplasia	DiGeorge syndrome	Primary tracheal tumour	Teratomas	Thymoma
Deletion of 12q	Cariofaciocutaneous syndrome	Fryn's syndrome	Goitre	Lymphatic malformation	Lymphoma
Brachmann–de Lange syndrome	Camptomelic dysplasia	De la Chapelle dysplasia	Neuroblastoma	Haemangiomas	Bronchogenic cysts
Pierre Robin syndrome	Crouzon syndrome	Noonans syndrome	Enterogenous cysts	Cystic hygromas	
Chitayat syndrome	Spondyloepiphyseal dysplasia congenital	Spondylocostal dysostosis			
Late-onset Pompe's disease	Loeys–Dietz syndrome	Filamin A mutation			
Osteogenesis imperfect	Hallermann–Streiff syndrome				

CHARGE: coloboma, heart anomaly, choanal atresia, retardation, genital and ear anomalies; VATER: vertebrae, anus, trachea, oesophagus and renal; PAH: pulmonary arterial hypertension; TOF: tracheo-oesophageal fistula.



ERS statement on tracheomalacia and bronchomalacia in children

Clinical spectrum in Tracheomalacia

Primitive
Isolated TM

TM secondary to Prematurity

Congenital Syndrome

Significant And Complex
Comorbidities

TM Secondary To Isolated AE/TEF



PRIMARY CARE PEDIATRICIAN

PAEDIATRICS OF COMPLEXITY

Courtesy of
S.Buratti
and A.Conte



Primitive Isolated TM

- Estimated to be the **most common form**, still under-diagnosed.
- Usually **autoresolving**, mild form with self-limited course (resolution by **2 years of age**) ⁽¹⁾
- May undergo **anterior aortopexy (AP)**.
 - 30% of patients who undergo AP ⁽²⁾
- Isolated bronchomalacia is a relatively rare condition ⁽²⁾
- Central role of the **Primary Care Pediatrician**






1. Choi S, et al .Diagnosis, Classification, and Management of Pediatric Tracheobronchomalacia: A Review. JAMA Otolaryngol Neck Surg. 2019 Mar 1;145(3):265.

2. Ghezzi M, et al. Airway Malacia: Clinical Features and Surgical Related Issues, a Ten-Year Experience from a Tertiary Pediatric Hospital. Children. 2021 Jul 20;8(7):613.

Primitive Isolated TM

The role of the pediatrician in caring for children with tracheobronchomalacia

Manisha Ramphul^a, Andrew Bush ^{b,c}, Anne Chang ^{d,e}, Kostas N Priftis ^f, Colin Wallis^g, Jayesh Mahendra Bhatt ^{h*}
and On behalf of the European Respiratory Society Task Force on tracheobronchomalacia in children



6.3. Optimizing general respiratory health

As with the management of other long-term respiratory conditions, the negative impacts of secondhand hand smoking or vaping should be considered [71–73]. Children should also receive their scheduled immunizations, as well as the annual influenza vaccination. Dry warm housing is beneficial [74] and the positive value of physical exercise should not be discounted.

6.4. The role of airway clearance therapy (ACT)

There is a paucity of evidence for the use of ACT in TBM.

7. Management of comorbidities

7.1. Gastro-oesophageal reflux (GOR)

7.2. Eosinophilic esophagitis

9. Providing support to the child and the family

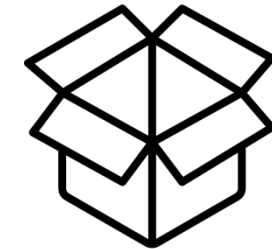
The pediatrician's role in the management of TBM includes providing support to the child and their family [99]. In order to be able to do so, the pediatrician should be able to understand the child's and parents' beliefs and values [11]. For

The role of the pediatrician is to tailor a management plan for the individual child, which as far as is possible, is evidence-based. Once the diagnosis is made, the pediatrician will be in a position to discontinue any unnecessary prior treatment



Bundle for the Primary Care Pediatrician

GENERAL	Longitudinal growth assessment
RESPIRATORY	Aerosolic therapy (avoid β -agonist) Barking cough Wheezing Progression/recurrence of symptoms ALTE – BRUE
GASTROINTESTINAL	Prevention of GERD Suspicion of eosinophilic esophagitis
INFECTIOUS	Recurrent pneumoniae Protracted bacterial bronchitis (PBB) Rule out primitive immunodeficiency
POST SURGICAL	Follow up for surgery related complications Scoliosis
Providing support and information to the child and their family	



Choi S, Lawlor C, Rahbar R, Jennings R. Diagnosis, Classification, and Management of Pediatric Tracheobronchomalacia: A Review. JAMA Otolaryngol Neck Surg. 2019 Mar
Wallis C, Alexopoulou E, Antón-Pacheco JL, Bhatt JM, Bush A, Chang AB, et al. ERS statement on tracheomalacia and bronchomalacia in children. Eur Respir J. 2019 Sep
Ramphul M, et al. The role of the pediatrician in caring for children with tracheobronchomalacia. Expert Rev Respir Med. 2020 Jul 2;14(7):679–89

*Courtesy of
S. Buratti
and A. Conte*

TMass associated with prematurity

Deposit of cartilage starts at 7^o week of gestation
until about 25 weeks' gestation ⁽¹³⁾

Prevalence of TBM in BPD is estimated to be **10-48%** ⁽¹³⁾
48-60% in severe forms ⁽¹⁴⁾

Infants with TBM have **>60% frequency of rehospitalization**
during 1^o year of life⁽¹³⁾

For severe forms: **NIV** (CPAP) as main treatment or need for
tracheostomy due to diffuse dynamic collapse ⁽¹⁴⁾

In extreme preterm, tracheomegaly can occur ⁽¹³⁾



Prima Edizione: 2022

<https://sinpia.eu/wp-content/uploads/2023/10/Il-followup-del-neonato-pretermine.pdf>

13. Hysinger EB. Central airway issues in bronchopulmonary dysplasia. *Pediatr Pulmonol.* 2021 Nov;56(11):3518–26.

14. Bush D, Juliano C, Bowler S, Tiozzo C. Development and Disorders of the Airway in Bronchopulmonary Dysplasia. *Children.* 2023 Jun 29;10(7):1127.

TM associated with congenital syndromes

Disease	Association with TBM anomalies	Ref.
Mucopolysaccharidosis	50-55% of patients with tracheal deformity	10.3390/diagnostics10010037
VACTERL	As per AE/TOF (50-80% of patients)	10.1186/1750-1172-6-56
CHARGE	Up to 40% of patients with 29% rate of tracheostomy	10.1001/archotol.125.1.33
Down syndrome	Depending on clinical symptoms	10.1002/ppul.25203
Achondroplasia	5.5% of airway malacia (↑ lower involvement)	10.1002/ajmg.a.36303
Neurofibromatosis	Incidence based on single reports, tracheal involvement by neurofibromas	10.1016/s0003-4975(02)03958-9

TABLE 1 Reported prevalence of tracheal anomalies in DS and general population

	DS (%)	DS with airway complaints and/or tracheobronchoscopy (%)
Congenital tracheal stenosis	0.4	2.8-5
Acquired tracheal stenosis	0.8	6
Complete tracheal ring deformity	-	4
Tracheal bronchus	-	3.1-21
Aberrant right subclavian artery	17-28.3	6
Tracheomalacia	4.4-7.1	23.1-51.5
Tracheal web	-	4

Abbreviations: DS, Down syndrome; -, not reported.

Adapted by Fockens et al

CRITICAL ROLE OF CLINICAL SUSPICION



Courtesy of
S. Buratti
and A. Conte

TMass associated with EA/TEF

Severe TBM is present in up to 11-33% of patients with corrected EA/TEF (1)



ERNICA Consensus Conference on the Management of Patients with Esophageal Atresia and Tracheoesophageal Fistula: Follow-up and Framework

Carmen Dingemann¹ Simon Eaton² Gunnar Aksnes³ Pietro Bagolan⁴ Kate M. Cross⁵
 Paolo De Coppi^{5,6} JoAnne Frulthof⁷ Piergiorgio Gamba⁸ Steffen Husby⁹ Antti Koivusalo¹⁰
 Lars Rasmussen¹¹ Rony Sfeir¹² Graham Slater¹³ Jan F. Svensson¹⁴ David C. Van der Zee¹⁵
 Lucas M. Wessel¹⁶ Anke Widenmann-Grolig¹⁷ Rene Wijnen¹⁸ Benno M. Ure¹

Table 1 Consensus statements on the follow-up of patients with esophageal atresia and tracheoesophageal fistula

N°	Statement	ESPGHAN-NASPGHAN Statement available ⁸	Consensus	%	Votes	Median (range)
1	There should be a structured schedule for life-long follow-up.	-	+	100	18/18	9 (8-9)
2	There should be an interdisciplinary follow-up program including surgery, gastroenterology, pulmonary, nutrition counselling, otolaryngology, and others, with one specialist leading.	+	+	100	18/18	9 (8-9)

Gaslini

 ISTITUTO GIANNINA GASLINI ISTITUTO PEDIATRICO DI RIVIGLIANO & CLINICA A CARATTERE SCIENTIFICO	Percorso nascita e trattamento del neonato affetto atresia esofagea	 Joint Commission International
	PDTA	Pagina 1 di 36
		PDTA-DIPCHIR-002

Feb 2019

Courtesy of
 S. Buratti
 and A. Conte



TM associated with EA/TEF

Evaluation	0-6 months	6-12 months	1-6 years	>6 years
Respiratory				
Clinical assessment	Every 3 months	Every 3 months	Every 6 months	Every 12-24 months
Endoscopy (FBS)	Clinical indication	At 6 months	Clinical indication	
Lung function tests	n/a		At 6 years	Every 12-24 months
Angio-CT, bronchoscopy, aortopexy (...)	Clinical indication			
Gastroenterological				
Clinical assessment	Every 3 months	Every 3 months	Every 6 months	Every 12-24 months
Impedance-pH monitoring	Clinical indication	At 12 months (after PPI discontinuation)	Clinical indication	
EGDS	Clinical indication	At 12 months (after PPI discontinuation)	Clinical indication	At 10 years (before transition to adult-care)
Surgical				
Clinical assessment	In the first three months	According to clinical indication		
Neurological				
Psychometric evaluation	Regular follow up at least until 3 years			Pre-scholar evaluation
Cardiac				
Clinical + Echocardiography	If necessary			
Speech therapy assessment				
Feeding strategies	Regular follow up according to personal needs			
Video-fluoroscopy	If necessary			
Orthopaedic				
Clinical + Rx assessment	At 12 months		If necessary	



Future perspectives – Radiation sparing follow-up

TABLE 2 Suggested diagnostic tests for patients with TEF according to their age

Test	Infant	Child	Adolescent
Bronchoscopy (flexible & rigid)	Yes	prn	prn
Pulmonary function testing	(?) ^a	Yes	Yes
Modified barium swallow/ videofluoroscopy/FEES study	Yes ^b	prn ^c	prn ^c
Chest X-ray	Yes	prn	prn
Chest CT	(?) ^d	Yes ^f	Yes ^f
Upper-GI endoscopy	Yes	prn	prn
Echocardiogram	Yes	prn	prn

Abbreviations: CT, computed tomography; FEES, fiberoptic endoscopic evaluation of swallowing; GI, gastrointestinal; TEF, tracheoesophageal fistula.

^aModerate or weak support because equipment are not readily available everywhere.

^bbefore oral feedings are introduced; prn

^cfor suspected aspiration.

^dmultidetector CT for evaluation of tracheomalacia & with contrast for vascular abnormalities.

^ffor detection and/or follow-up of bronchiectasis.

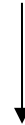


TM associated with significant and complex comorbidities



ERS statement on tracheomalacia and bronchomalacia in children

Pragmatically, it may also be helpful to distinguish conditions where TBM is clinically the main problem and those in which, while still a factor, there are either more important complex extrapulmonary comorbidities such as cardiovascular abnormalities or pulmonary parenchymal disease.



NEED FOR MULTIMODAL CARE

A cartoon illustration of a man with a large red turban and a blue coat with a white collar. He has a mustache and is looking towards the right with a surprised expression. He is holding a white skull in his right hand. The background is a light beige color with a large, faint thought bubble shape behind him.

**Follow-up del paziente
con tracheomalacia:
indicazione ad intervento
chirurgico?**

Questionario Clinico nei pazienti con tracheomalacia

01

0-2 years:

ALTEs; persistent or recurrent barking cough; **in- or expiratory stridor**; increased work of breathing; **recurrent and prolonged respiratory infections affecting the lower tract.**

02

2-6 years: persistent or

recurrent barking cough; recurrent and prolonged respiratory infections of the lower tract (>6/year); **poor resistance to play**; in- or expiratory stridor, dysphagia or recurrent vomiting.

03

>6 years: persistent or

recurrent barking cough; exercise intolerance; **cough under stress, not accompanied by bronchospasm**; recurrent and prolonged respiratory infections affecting the lower tract (>6/year); **dysphagia or vomiting and/or GER.**

Ruolo dell'endoscopia

0 point: < 50 % reduction

1 point: mild (50–75% reduction)

2 points: moderate (75–90% reduction)

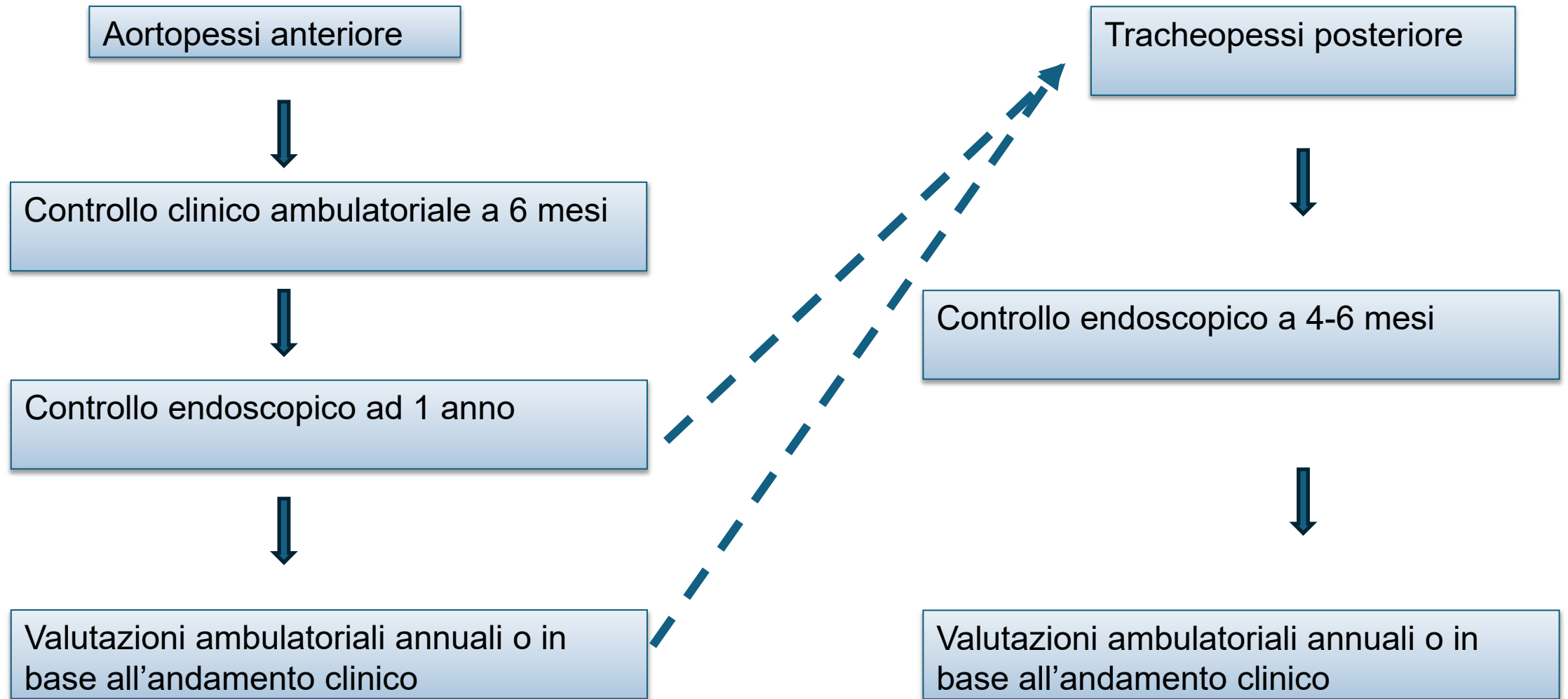
3 points: severe (>90% reduction)

Team Vie Aeree

Valutazione multidisciplinare

- Anestesisti e rianimatori
- Cardiochirurghi
- Chirurghi toracici
- Gastroenterologi
- Otorinolaringoiatri
- Pneumologi
- Radiologi
- GENITORI

Tracheomalacia s/p ad Intervento chirurgico



Tracheomalacia NON s/p ad intervento chirurgico

Valutazioni ambulatoriali annuali (clinica e PFR) o in base all'andamento clinico



Rivalutazione globale (FBS ed eventuale ripetizione angioTC) se peggioramento clinico e funzionale

F, 16 anni

Trattata per Asma senza beneficio

>6 years:

**persistent or recurrent
barking cough;**

exercise intolerance;

**cough under stress, not
accompanied by
bronchospasm;**

recurrent and prolonged
respiratory infections affecting
the lower tract (>6/year)

Score clinico: 4

Score endoscopico: 3



F, 16 anni

Controllo endoscopico post-aortopessi

>6 years:

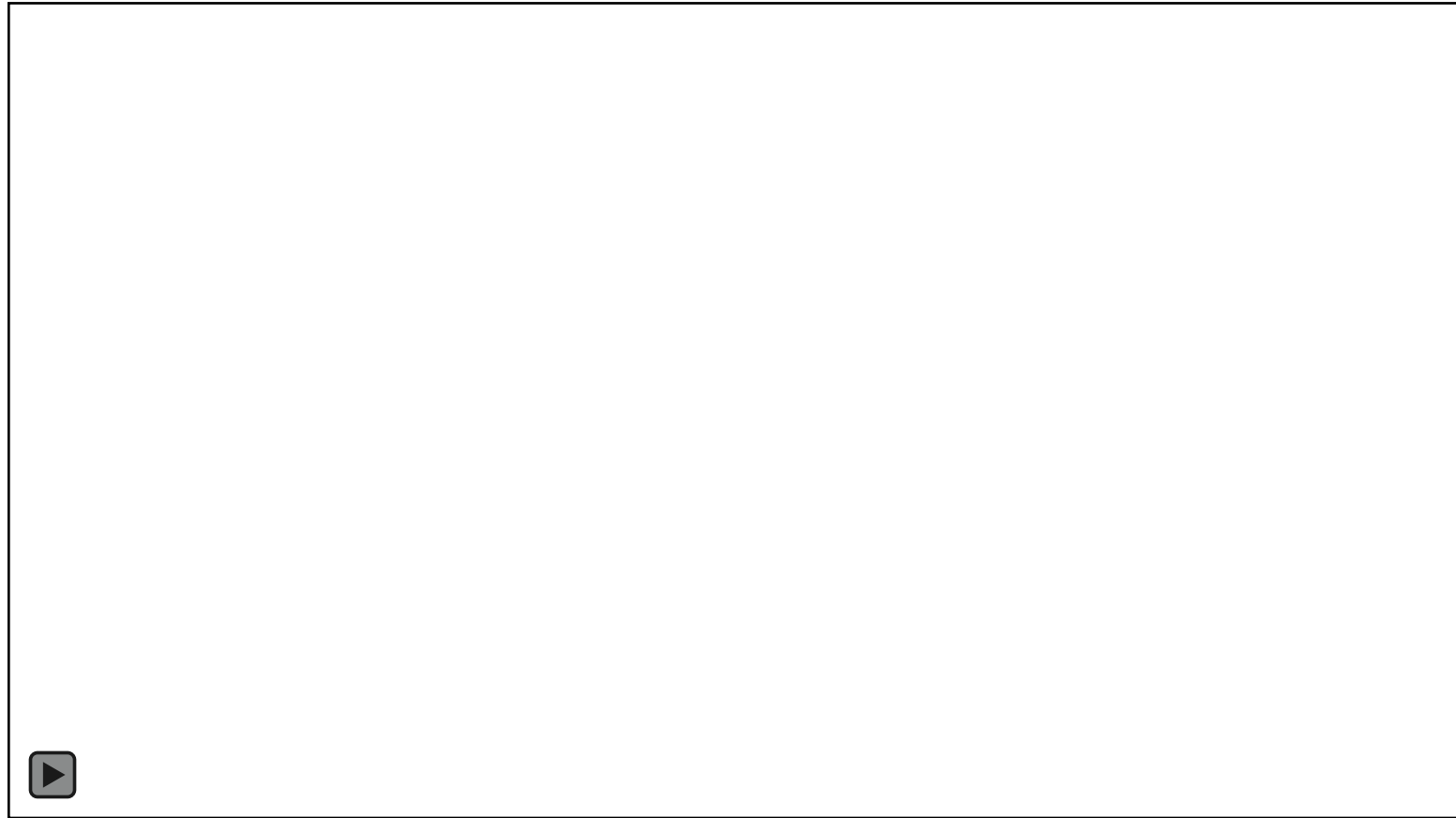
**persistent or recurrent
barking cough;**

exercise intolerance;

**cough under stress, not
accompanied by
bronchospasm;**

Score clinico: 3

Score endoscopico: 2



F, 16 anni

Controllo endoscopico post-tracheopessi

>6 years:

**cough under stress, not
accompanied by
bronchospasm;**

Score clinico: 1

Score endoscopico: 0



SURGICAL TREATMENT OF TRACHEOBRONCHOMALACIA IN CHILDREN: INDICATIONS, TECHNIQUES, AND

RESULTS *Exploratory data F. Lena, M. Torre et al UOC di Chirurgia Pediatrica IGG*

Patients characteristics stratified by age

Characteristics	Total Cohort (N= 143)	0-2 years (N= 43)	2-6 years (N= 63)	≥ 6 years (N= 37)
	n (%) or median±SD (IQR)	n (%) or median±SD (IQR)	n (%) or median±SD (IQR)	n (%) or median±SD (IQR)
Demographic				
Male	88 (61.5%)	28 (65.1%)	39 (61.9%)	21 (56.8%)
Age at 1 st surgery (months)	68±56 (2-278)			
Patient History				
AVC	99 (69.2%)	20 (46.5%)	48 (76.2%)	31 (83.8%)
EA/TEF	41 (28.7%)	22 (51.2%)	13 (20.6%)	6 (16.2%)
Other TM causes	3 (2.1%)	1 (2.3%)	2 (3.2%)	-
Clinical Score				
1 point	34 (23.8%)	13 (30.2%)	13 (20.6%)	8 (21.6%)
2 points	83 (58%)	23 (53.5%)	38 (60.3%)	22 (59.5%)
3 points	25 (17.5%)	7 (16.3%)	12 (19.1%)	6 (16.2%)
4 points	1 (0.7%)	-	-	1 (2.7%)

TM= tracheomalacia, EA/TEF=esophageal atresia/tracheoesophageal fistula, AVC= anteriorvascularcompression

Details of surgical procedures

Parameters	AA (N= 132)	PT (N= 18)	P value
	n (%) or median±SD (IQR)	n (%) or median±SD (IQR)	n
Weight at surgery (Kg)	17±15 (4-60)	51±21 (15-67)	< 0.001
Age at surgery (months)	55±49 (2-200)	125±68 (41-279)	< 0.001
Hospital stay (days)	8±7 (4-50)	8±11 (3-16)	0.4956
Complications			
• Overall	25 (18.9%)	3 (16.7%)	> 0.99
• Grade ≥3	3 (2.3%)	1 (5.6%)	0.4036

Post-operative outcomes

	Pre-op Cohort (N= 143)	Follow-up Population (N= 116)	P value
	n (%) or median±SD (IQR)	n (%) or median±SD (IQR)	n
Clinical score			
0 points	0 (0%)	68 (58.6%)	p <0.001
1 point	34 (23.8%)	47 (40.5%)	p =0.0046
2 points	83 (58%)	10 (8.6%)	p <0.001
3 points	25 (17.5%)	1 (0.9%)	p <0.001
4 points	1 (0.7%)	0 (0%)	p >0.99

Courtesy of F. Lena
and M. Torre



Grazie dell'attenzione

