

Anomalie des Arcs Aortiques

A.Bonnard

Hôpital Robert Debré

Objectifs

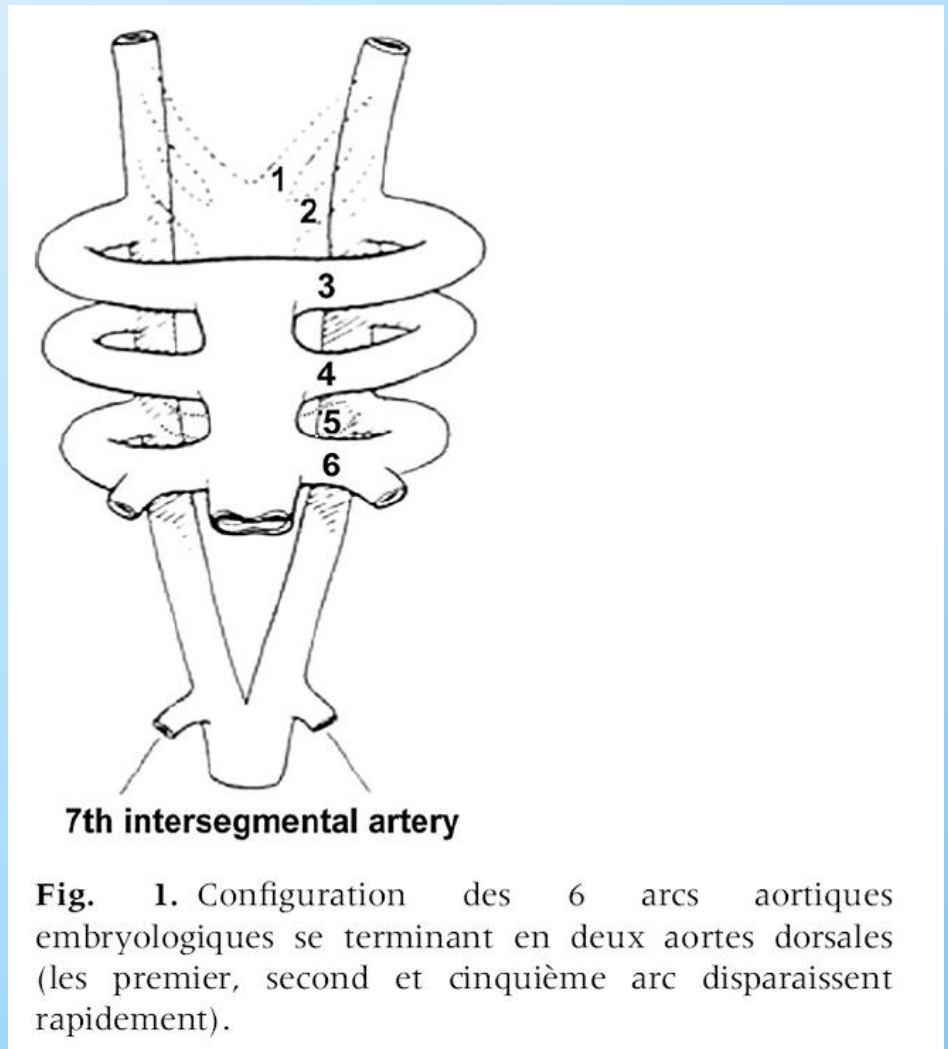
- Épidémiologie
- Diagnostic et malformations associées
- Prise en charge

Épidémiologie

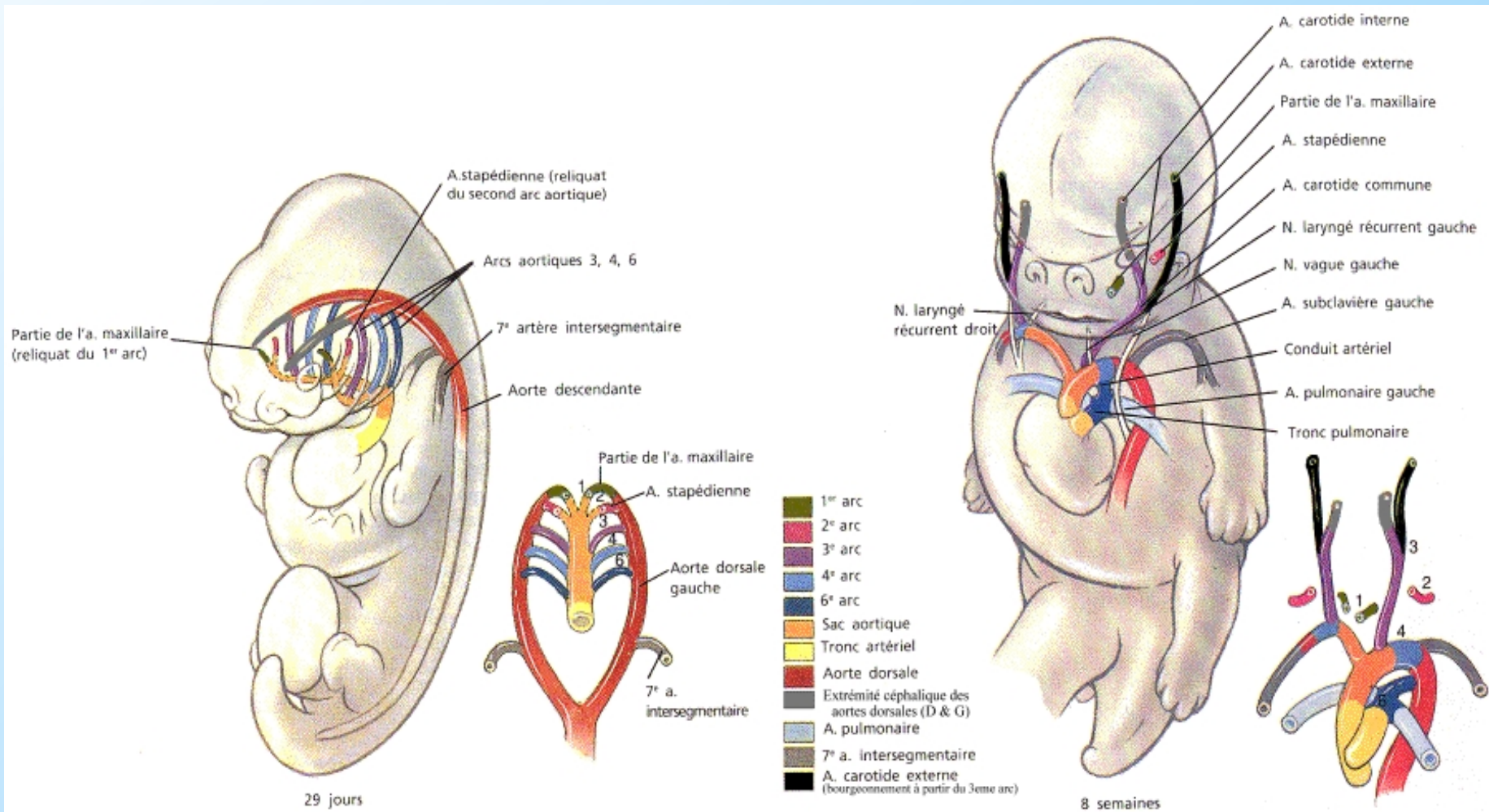
- <1% de toutes les défaillances cardiaques congénitales
- fréquence id. dans les 2 sexes
- Pas de prédominance géographique ou raciale

Embryologie

- Anomalie de développement des arcs pharyngés



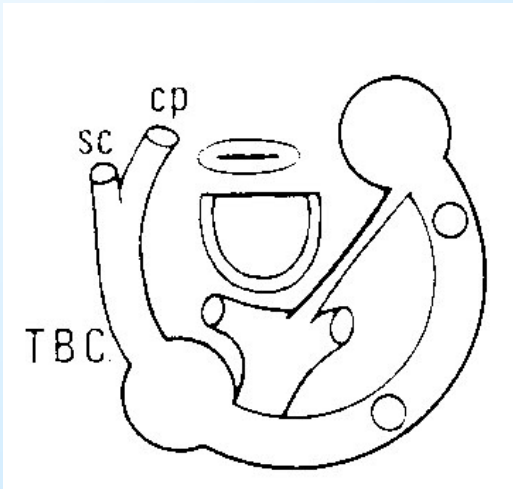
Embryologie



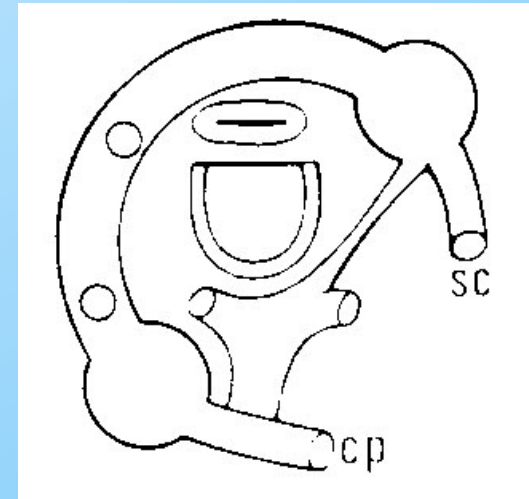
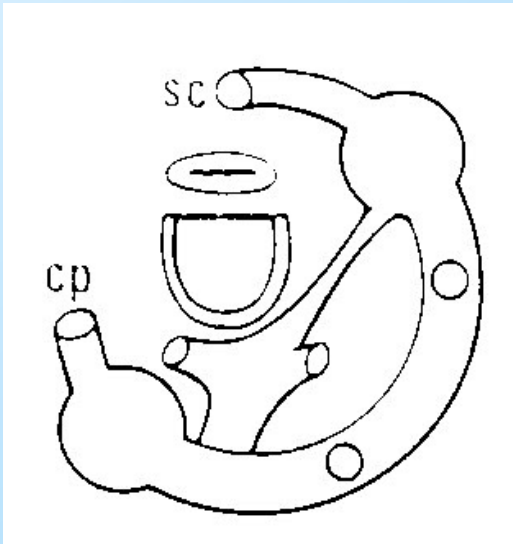
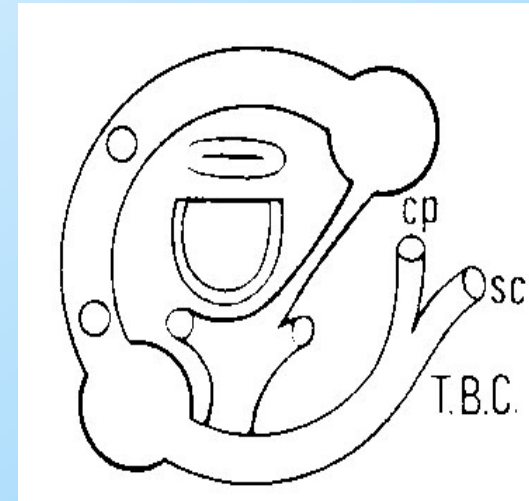
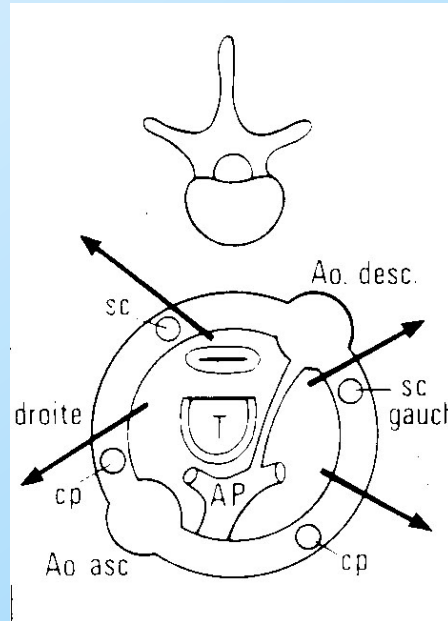
Embryologie

- gènes associés aux chromosomes 22Q11, tels que TBX1 et FGF8, semblent importants
 - Yamagishi H., Srivastava D. Unraveling the genetic and developmental mysteries of 22q11 deletion syndrome *Trends Mol Med* 2003 ; 9 : 383-389

Classification

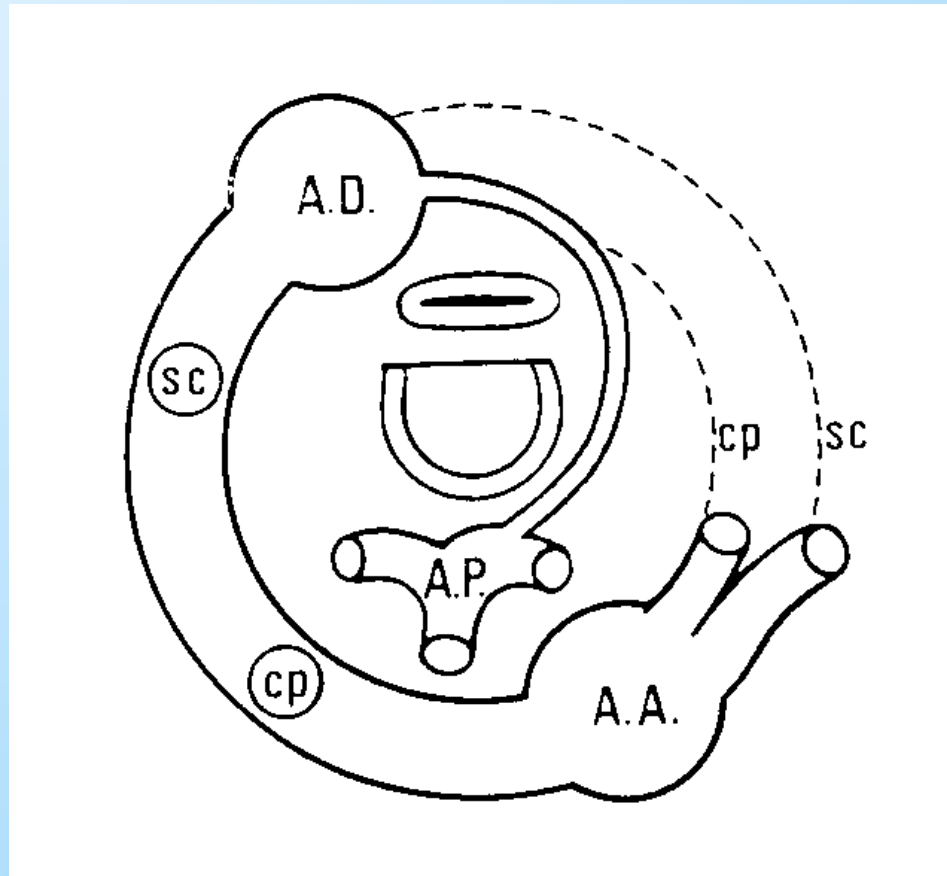


Aorte ascendante à droite

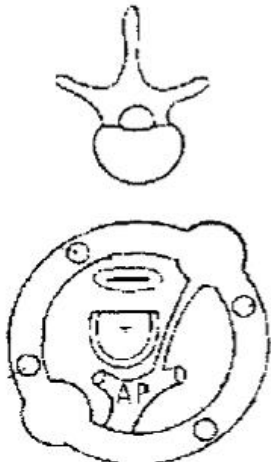
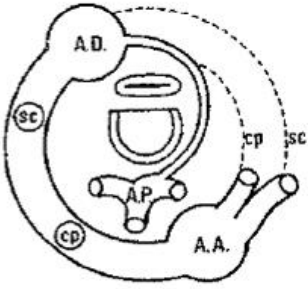
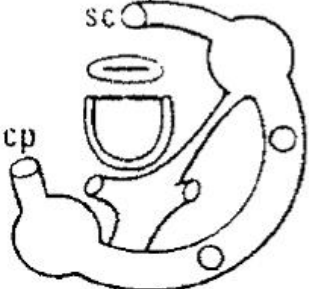
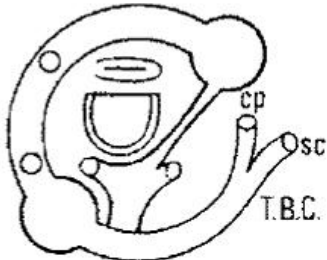


Classification

**Aorte ascendante
à gauche = anomalie de NEUHAUSER**



Classification

<p>COMPLETE DOUBLE ARCH 55%</p>  <p>The diagram shows a cross-section of the thoracic aorta with two arches. The upper arch is connected to the descending aorta by a single vessel. The lower arch is also connected to the descending aorta. The aortic arch is labeled 'A.P.'.</p>	<p>NEUHAUSER ANOMALY 22%</p>  <p>The diagram shows a cross-section of the thoracic aorta with a single arch. The arch is labeled 'A.D.' and is connected to the descending aorta by a vessel labeled 'A.A.'. The arch is also connected to the descending aorta by a vessel labeled 'sc'. The aortic arch is labeled 'A.P.'.</p>
<p>RIGHT ABERRANT SUBCLAVIAN ARTERY 16%</p>  <p>The diagram shows a cross-section of the thoracic aorta with a single arch. The arch is labeled 'sc' and is connected to the descending aorta by a vessel labeled 'cp'.</p>	<p>LEFT BRONCHIOCEPHALIC ARTERIAL TRUNC + LEFT DUCTUS ARTERIOSUS 7%</p>  <p>The diagram shows a cross-section of the thoracic aorta with a single arch. The arch is labeled 'cp' and is connected to the descending aorta by a vessel labeled 'sc'. The arch is also connected to the descending aorta by a vessel labeled 'T.B.C.'.</p>

Diagnostic

- Révélation

- Période néonatale: 28%
- 1ère année de vie: 68%
- >1an: 4%

96% < 1 an

- Anomalies associées: 15-20%

- **Chromosomique+++** : del 22q11 (DiGeorge, anomalies vélocardiofaciale), Rubinstein-Taybi (µdel 16p13.3)
- Cardiaque: PDA, CIV, CIA
- Digestif: AO
- Pulmonaire: emphysème

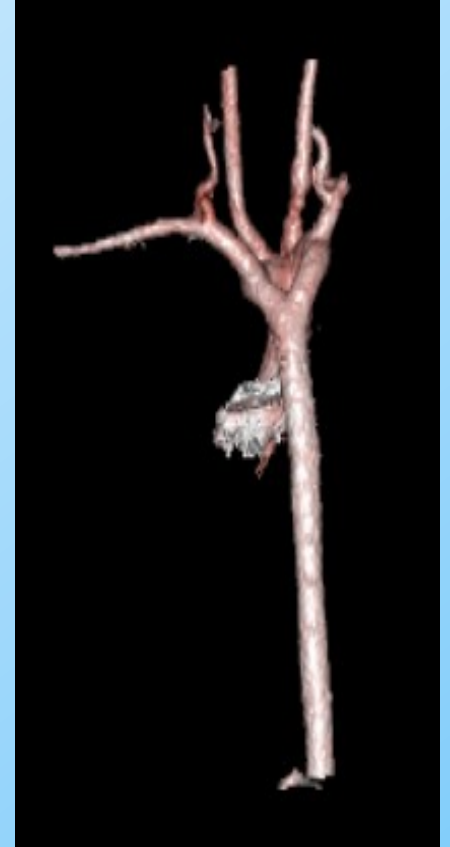
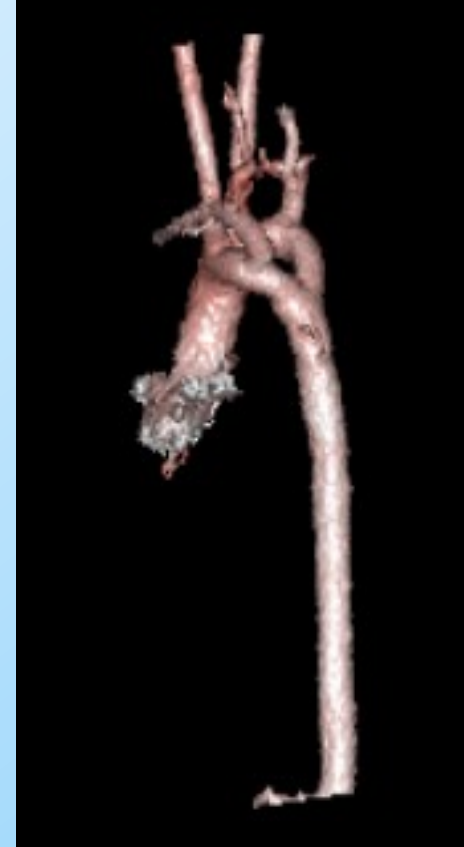
Diagnostic

- Clinique
 - **Respiratoire +++**
 - Détresse respiratoire néonatale
 - Stridor
 - Asthme du nourrisson
 - « Bronchiolite » à répétition
 - Pneumopathie à répétition
 - Digestif: dysphagie (dysphagia-lusoria), vomissements, RGO

Diagnostic

- Examens complémentaires
 - Échographie cardiaque
 - TDM, IRM
 - TOGD

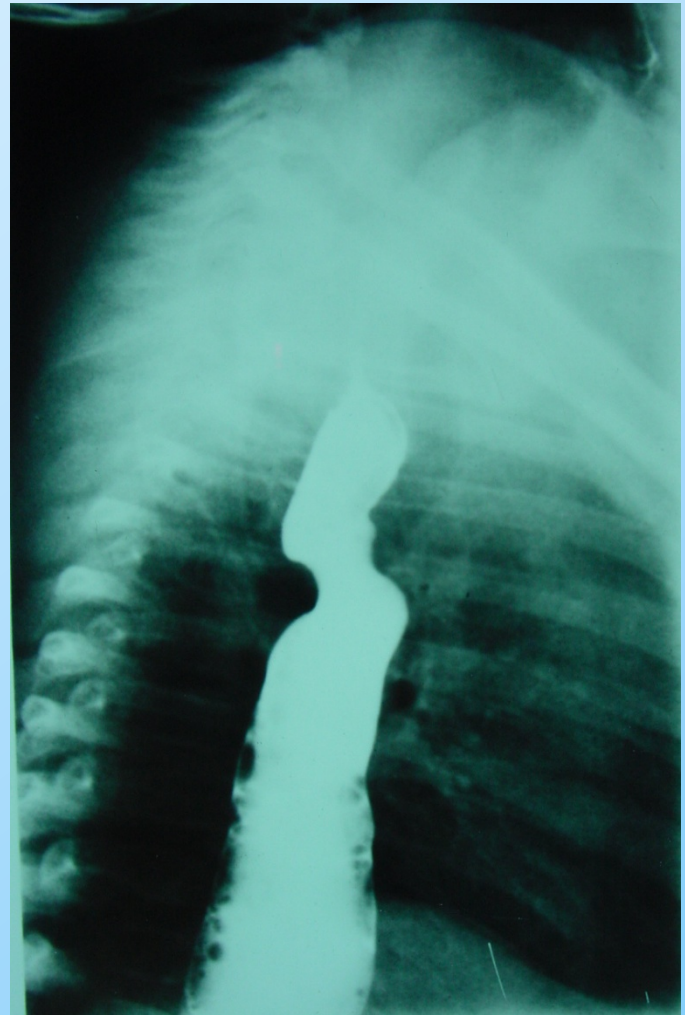
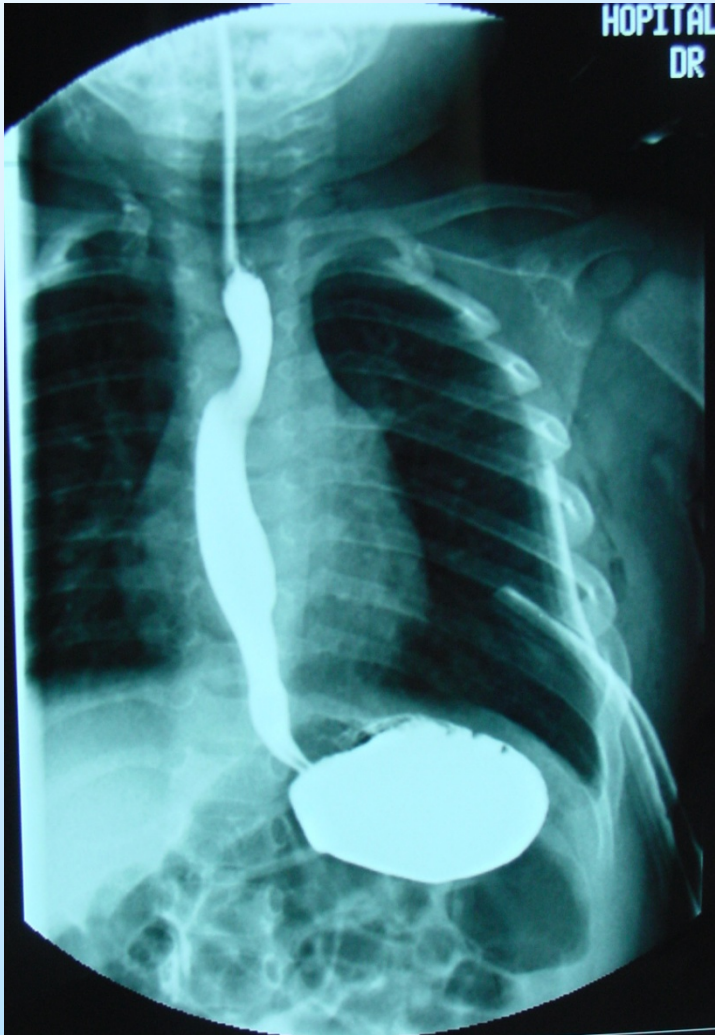
Diagnostic



Diagnostic



Diagnostic



Bilan

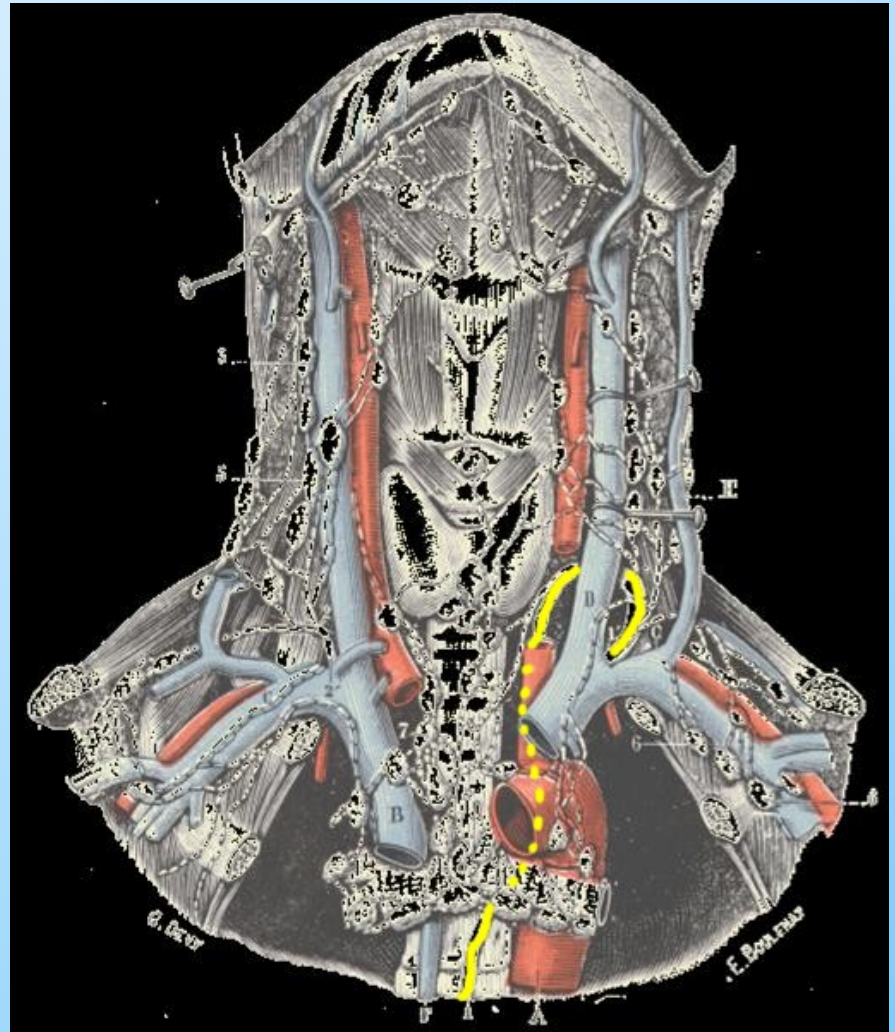
- Etiologique
 - Caryotype, recherche del 22q11
 - Consultation génétique si anomalies associées
- Retentissement
 - **Fibro ORL +++**
 - Compression trachéale (à évaluer)
 - Trachéomalacie
 - Signe indirect de RGO

Traitement

- Traitement médical
 - De la détresse respiratoire (ventilation invasive parfois nécessaire)
 - Du RGO
- Traitement chirurgical
 - Indication: symptomatique
 - Que dire aux parents?
 - Bénéfice de la chirurgie parfois retardé (trachéomalacie, RGO, maladie associée)
 - Risque: IRC, lésion trachéale irréversible, risque de la chirurgie

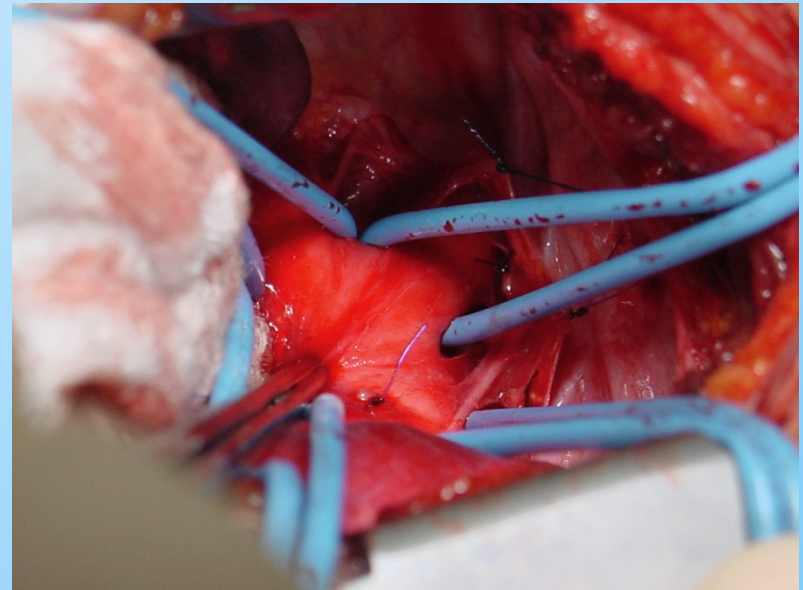
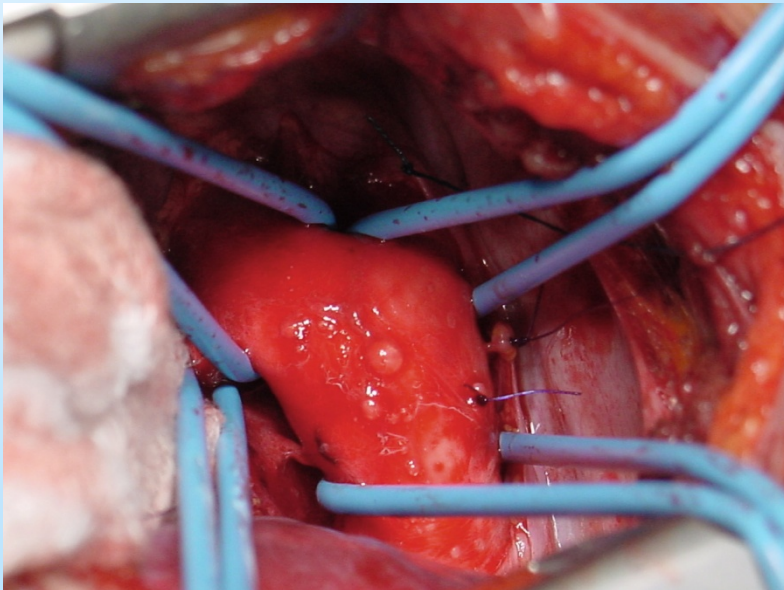
Traitement chirurgical

- **Voie d'abord**
 - Thoracotomie gauche ou thoracoscopie
 - Risques:
 - **Nerf:** X, phrénique, récurrent, canal thoracique
 - **Vaisseaux:** aorte, arc aortique
 - Œsophage, trachée



Traitement chirurgical

- Technique chirurgicale
 - **Identification parfaite des éléments anatomiques**
+++
 - Vasculaires (arc ant, arc post, sous clavière, aorte),
nerveux, œsophage, trachée



Traitement chirurgical

- Séparation du double arc de l'œsophage et de la trachée avant la section
- Ouverture de la fenêtre +++

15D.M.0131.00235268
07/06/2010
SI:4
< 208 - 4 Batch 3 (TOUT) >

Hopital R.DEBRE
CAR
22/06/2010, 10:34:47
PHILIPS

Résolution d'origine

LAF



FP

Complications

- **Per-opératoire**
 - Hémorragie
 - Plaie oesophagienne
 - Section du X
- **Post-opératoire**
 - Paralysie récurrentielle
 - Chylothorax
- **Persistance de la symptomatologie**
 - Par défaut de libération de l'œsophage et de la trachée
 - Trachéomalacie +++

Complications

Table 1. Factors Studied to Determine any Influence on Surgical Result

	Improvement			P Value
	Complete	Partial	No	
Age of revelation				
Neonatal 0-7 days	60%	30%	10%	Not significant
After	70%	15%	15%	
Associated anomalies				
Yes	67%	0%	33%	Not significant
No	67%	22%	11%	
Surgery				
<1 mo	43%	43%	14%	Not significant
>1 mo	74%	13%	13%	
Anomalies				
Complete double arch	53%	32%	15%	Not significant
Neuhauser	83%	0%	17%	
Right retro-oesophageal subclavian artery	100%	0%	0%	
Left bronchiocephalic arterial trunc and a left ductus arteriosus	100%	0%	0%	
Malacia				
Yes	50%	20%	30%	Not significant
No	83%	8.50%	8.50%	
Compression				
<50%	67%	22%	11%	Not significant
>50%	67%	0%	33%	

Conclusions

- Pathologie rare
- Y penser devant:
 - Détresse respiratoire néonatale sans étiologie évidente
 - Asthme sévère du nourrisson ou « bronchiolite » à répétition
- TDM
- Fibroscopie ORL pré opératoire indispensable