



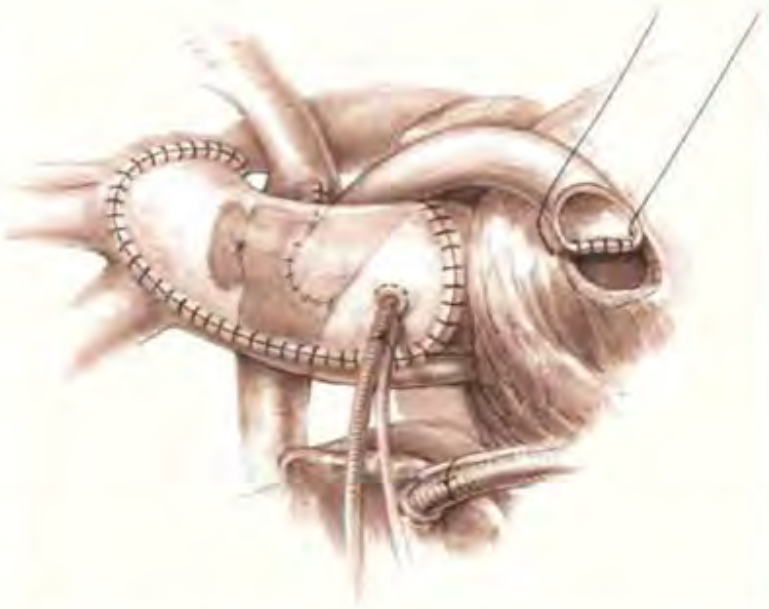
CHIRURGIE CARDIAQUE PEDIATRIQUE

DU de Cardiologie Pédiatrique

Vendredi 26 Janvier 2024

Dr Margaux PONTAILLER

COMPREHENSIVE
SURGICAL MANAGEMENT
OF CONGENITAL HEART DISEASES



RICHARD A. JONAS

ILLUSTRATED BY REBEKAH DODSON



Sommaire

Généralités

Techniques chirurgicales

Spécificités des GUCH

Assistance circulatoire

Transplantation

Cicatrices



Généralités



Voies d'abord

Deux grandes voies

- Sternotomie médiane
- Thoracotomie (droite ou gauche)

Avantages / inconvénients

- Types de cardiopathie / chirurgie
- Exposition
- Douleur
- Esthétique

D 651

AUTRES
DIRECTIONS

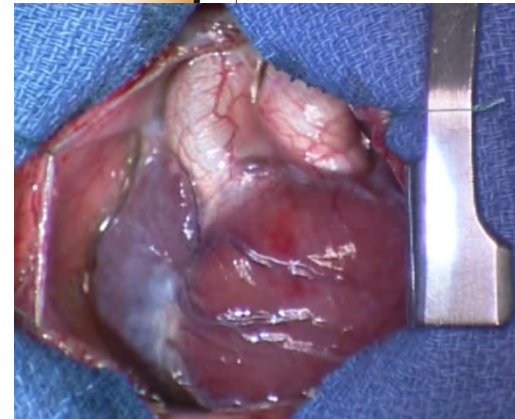
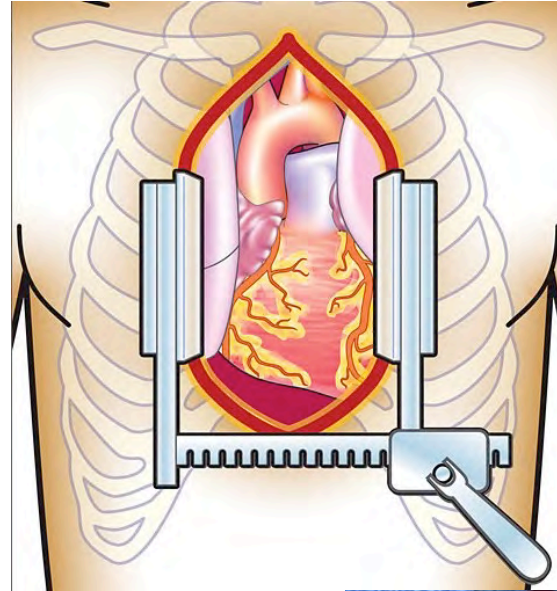
TOUTES
DIRECTIONS

Installation

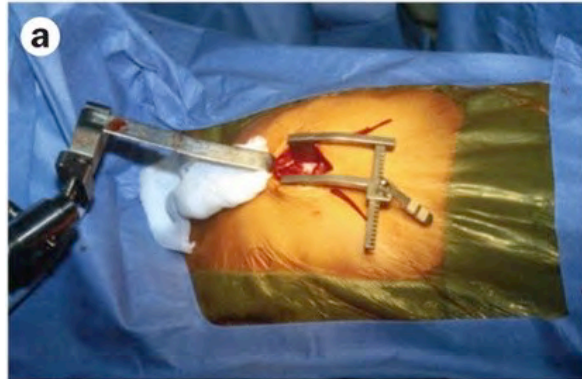
- Installation décubitus dorsal, billot sous les épaules
- Intubation oro ou naso trachéale
- Scope, saturomètre
- 1 à 2 VVP
- KTA
- KTC
- Sonde urinaire
- Sonde thermique
- Plaque de BE
- Plaques de défibrillateur externe si redux

Tout cela sur 0.2 m² de surface corporelle
=
Protection points d'appui

Sternotomie

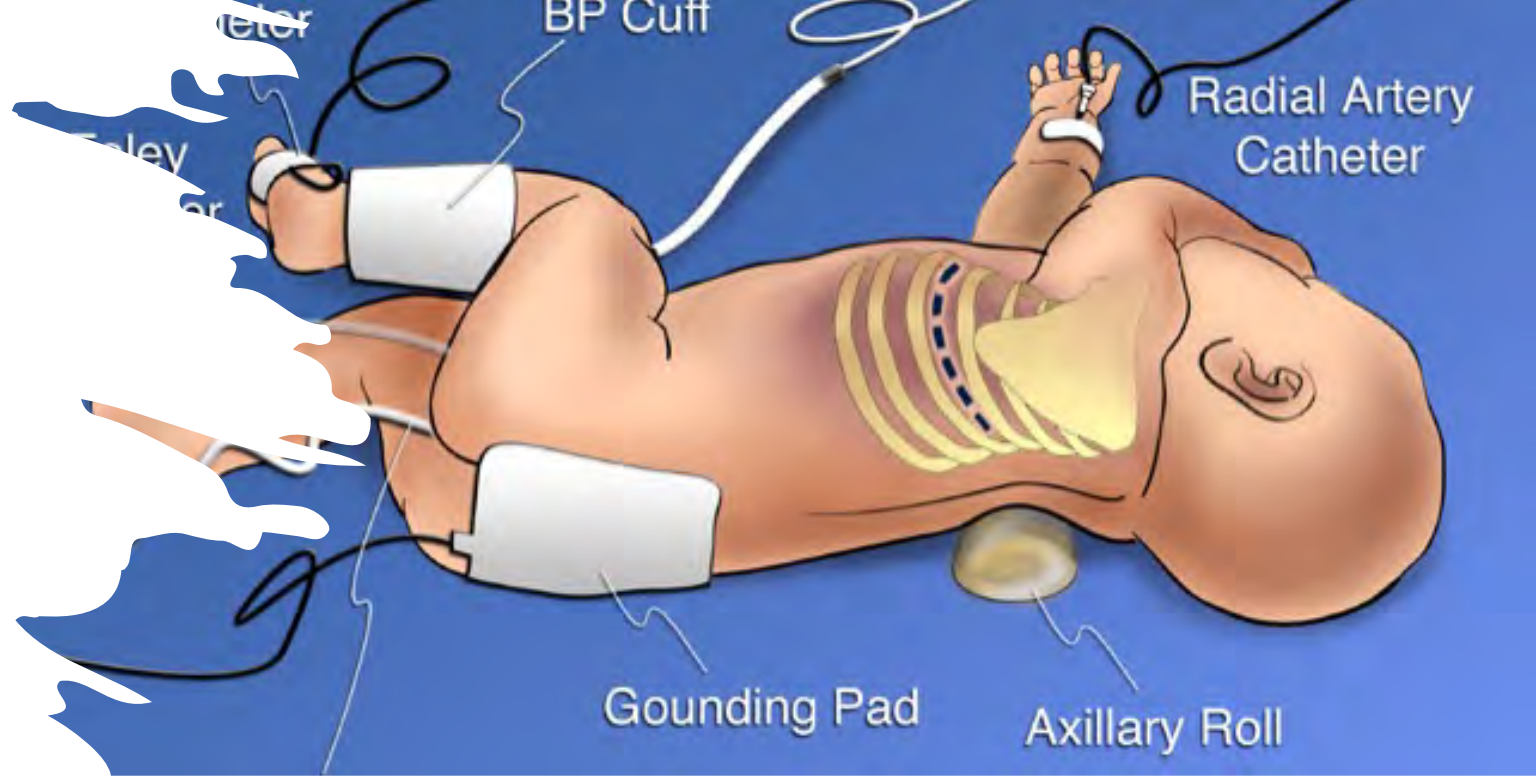


Sternotomie



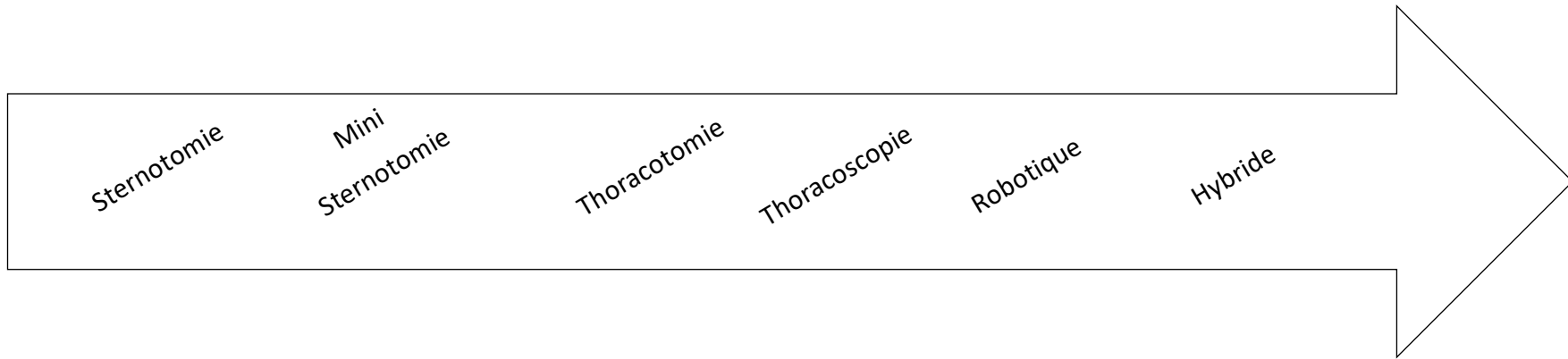
Thoracotomie

- Techniquage idem
- Installation en décubitus latéral D/G
- Main sur la tête
- Jambes en PLS
- Billot sous les côtes
- Protection points d'appui
+++

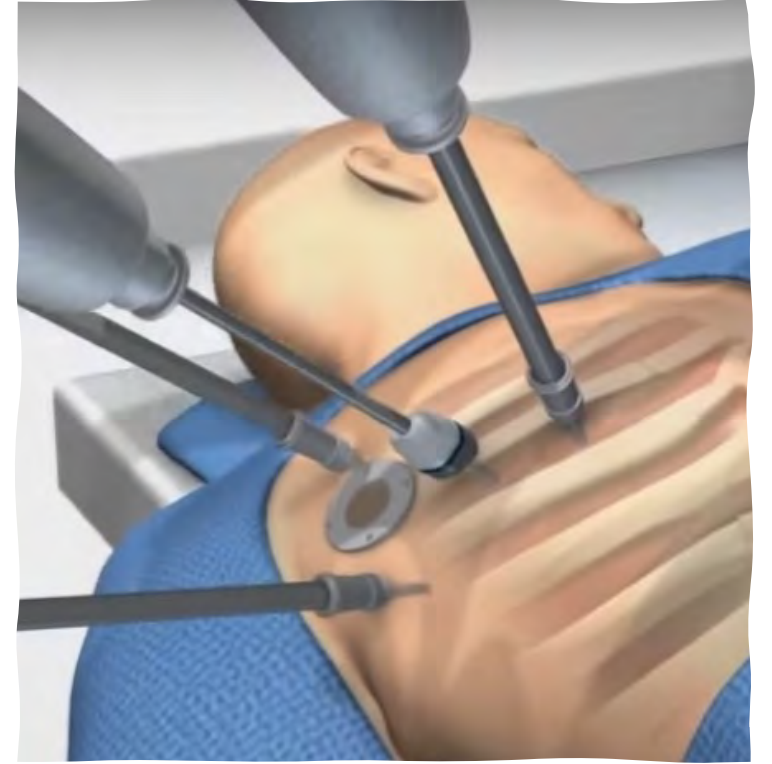




Thoracotomie



Chirurgie mini invasive



Chirurgie mini invasive

Principes de la chirurgie

Cœur fermé

versus

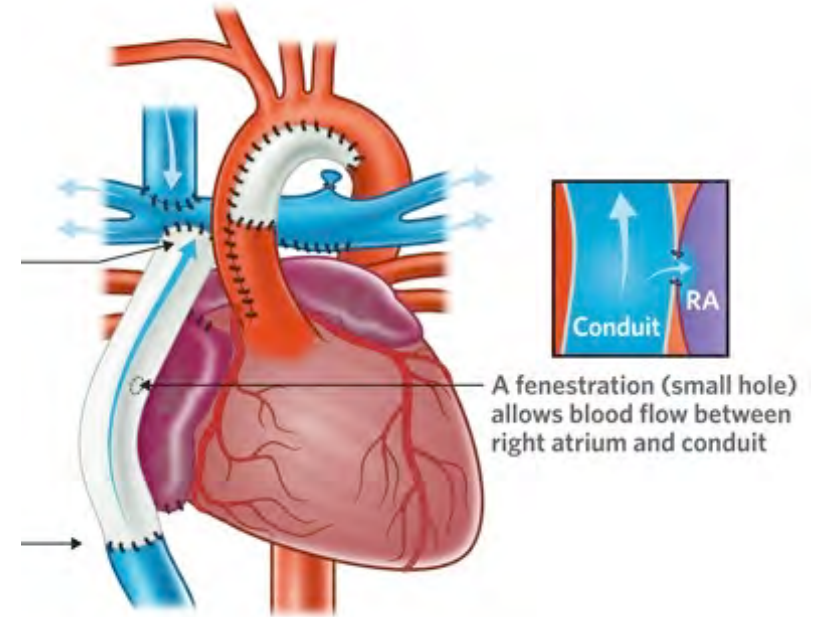
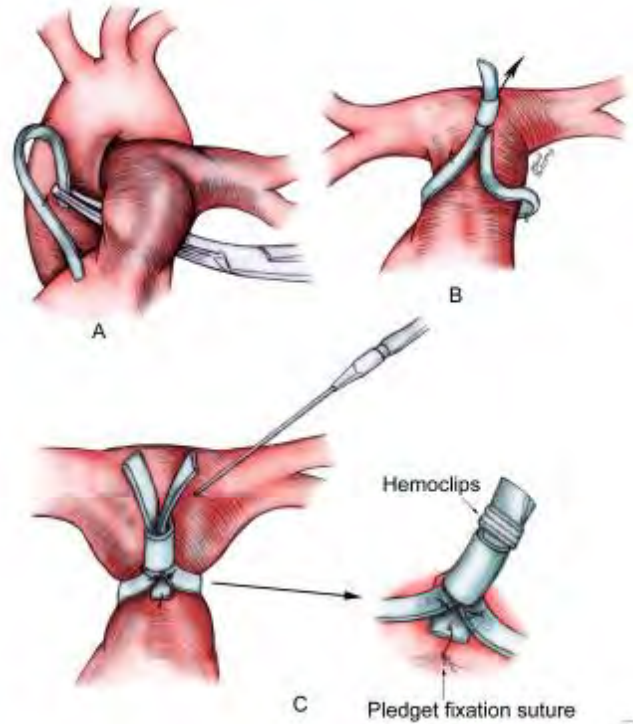
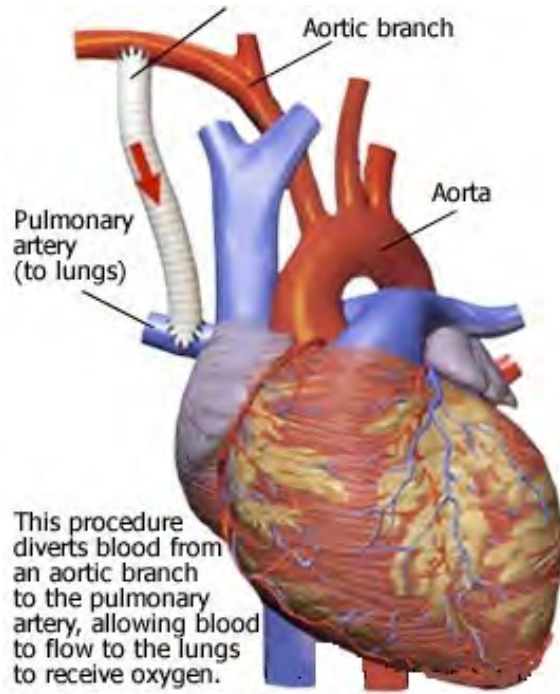
Cœur ouvert

Cure complète

Versus

Chirurgie palliative

Chirurgie palliative

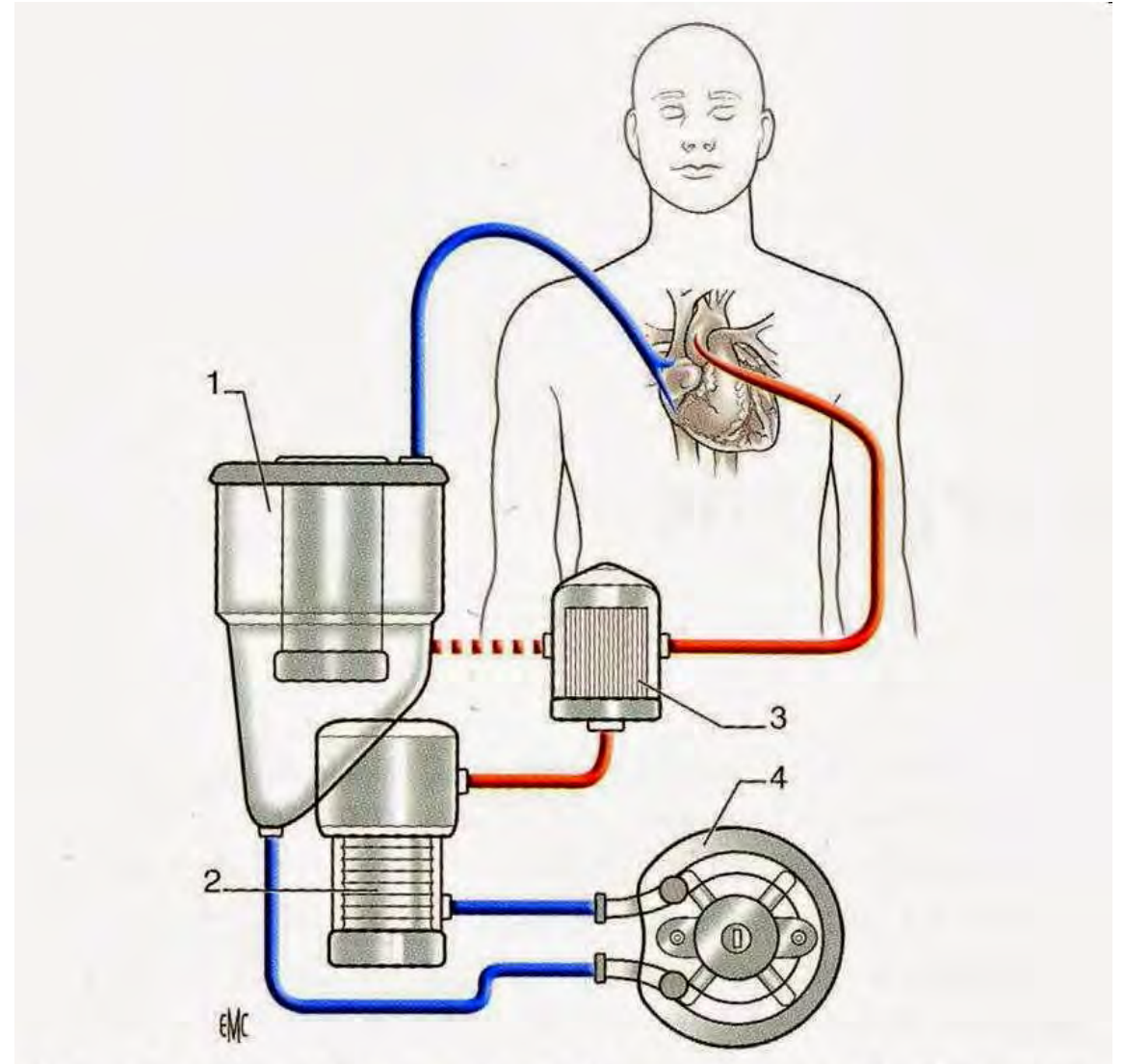


Coeur fermé = Pas de CEC

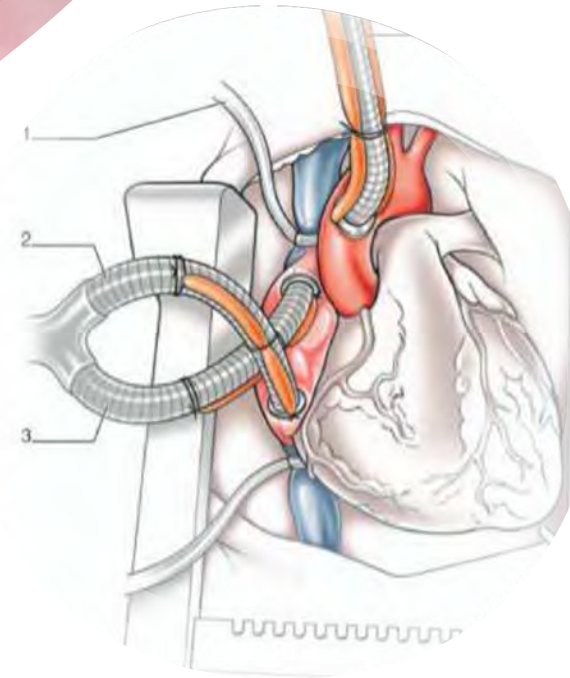
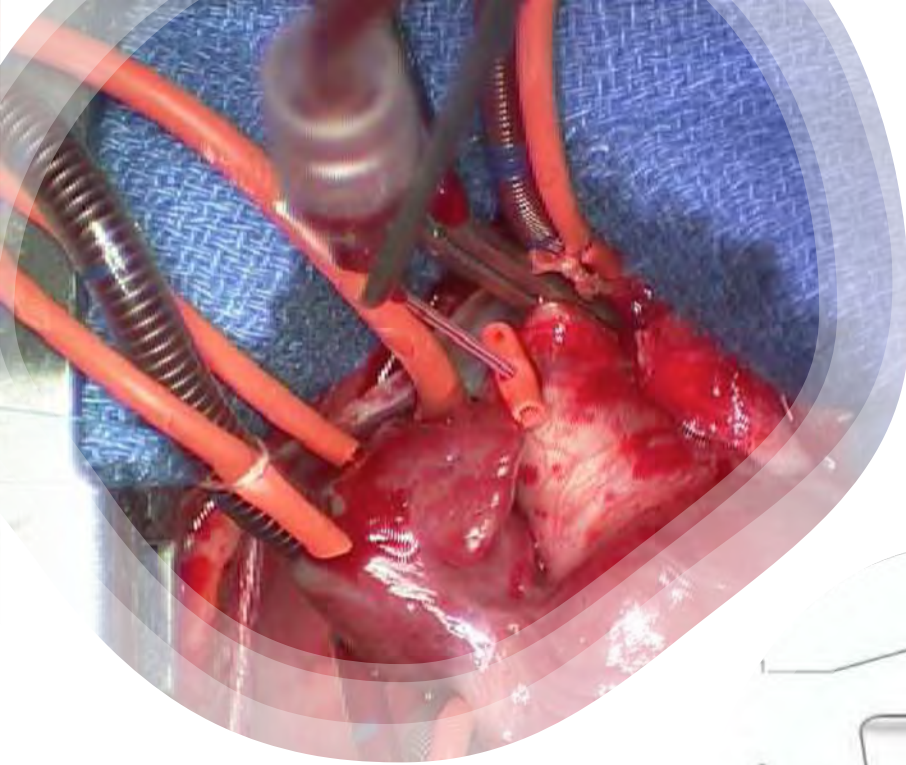
- Canal artériel
- Coarctation
- Pacemaker, défibrillateur
- Anastomose systémico-pulmonaire (Blalock)
- Anomalies arcs aortiques

Cœur ouvert = CEC

- Principes de la CEC
 - Assurer rôle du cœur
 - Assurer rôle du poumon
 - Visibilité des lésions
- Canules
 - Drainage
 - Réinjection
- Circuit
 - Réservoir
 - Filtre
 - Pompe
 - Oxygénateur
 - Echangeur thermique



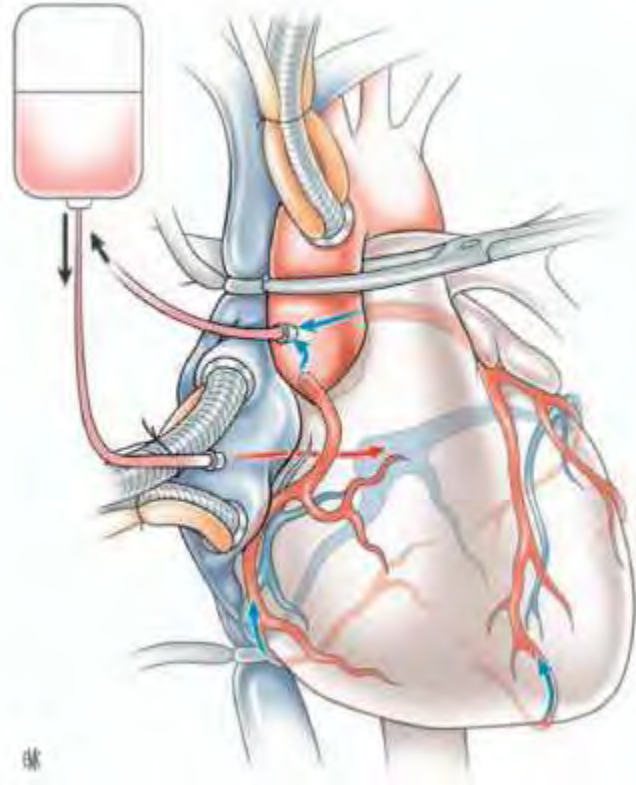
Canulation



- Canule artérielle
 - Ao ascendante : réinjection
- Canule(s) veineuse(s)
 - OD ou 2 veines caves : drainage
- +/- Canule de décharge gauche
 - OG : drainage
- +/- Canule de cardioplégie

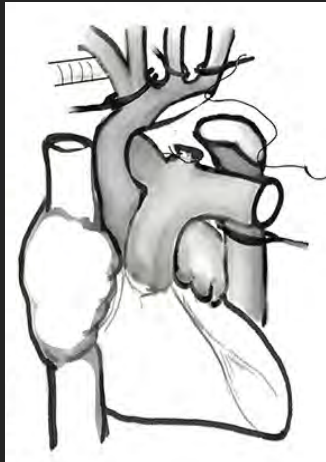
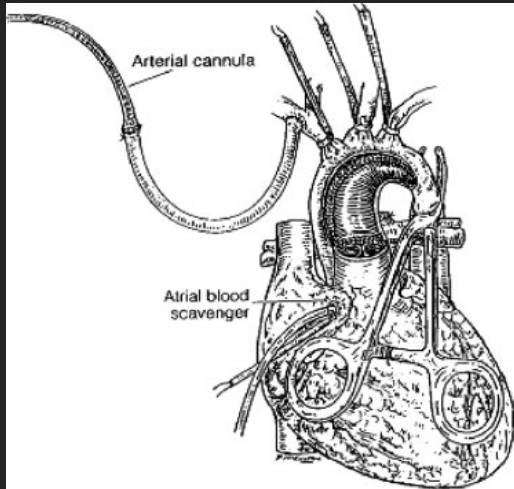
Cardioplégie

- Arrêter le cœur
- Perfuser le coeur



Cristalloïde / Sanguine
Chaude / Froide
Unique / Répétée / Continue
Antérograde / Rétrograde / Combinée

Normo ou hypothermie



- Normothermie
- Hypothermie modérée
- Hypothermie profonde
- Arrêt circulatoire
- Perfusion cérébrale sélective

Risques de la CEC



Hémodilution

Priming

Transfusion



Inflammation

Durée de CEC

Matériel



Ischémie myocardique

Cardioplégie

Temps de clampage



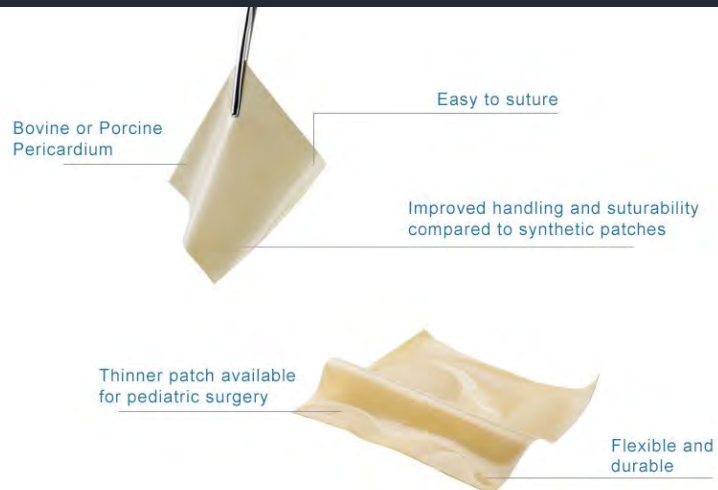
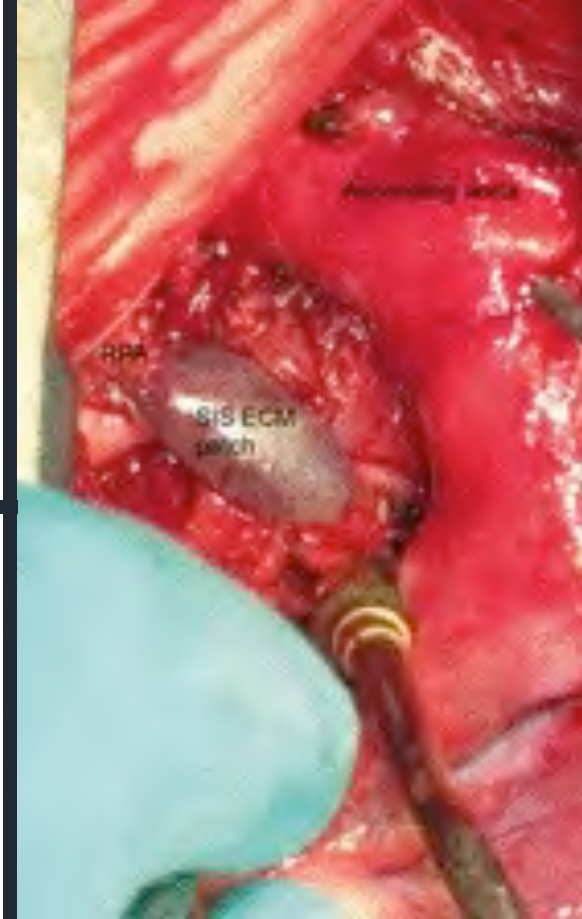
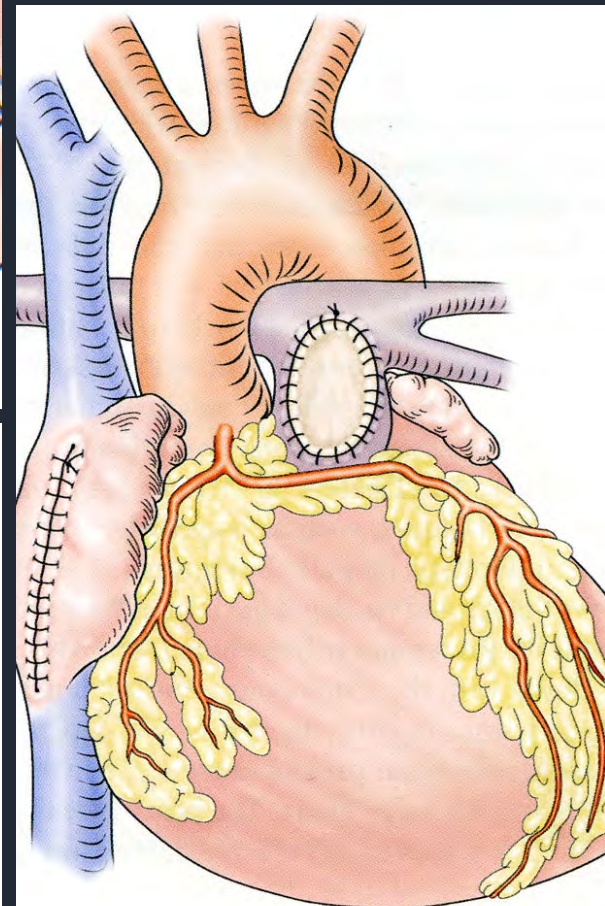
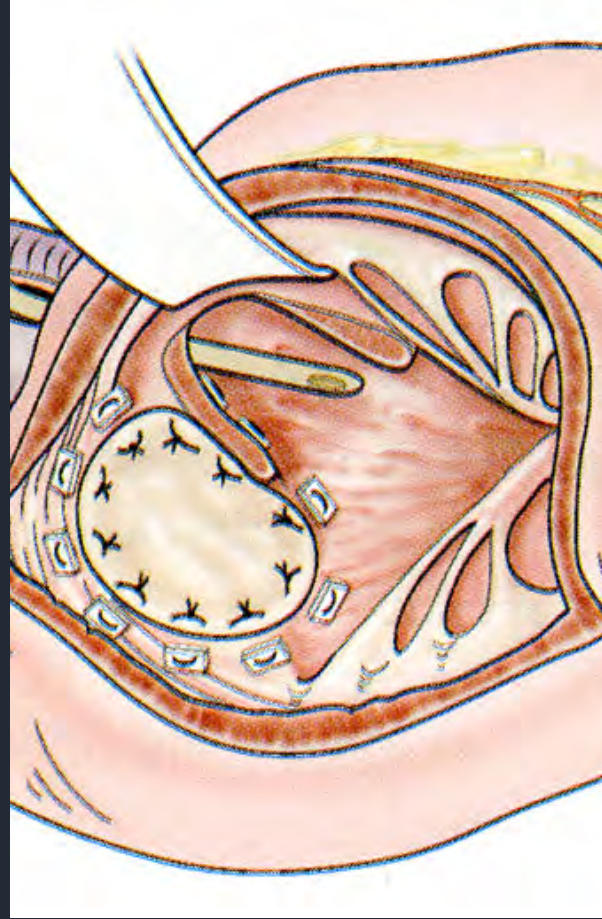
Problème technique

Décanulation

Bullage



Patches



Valves

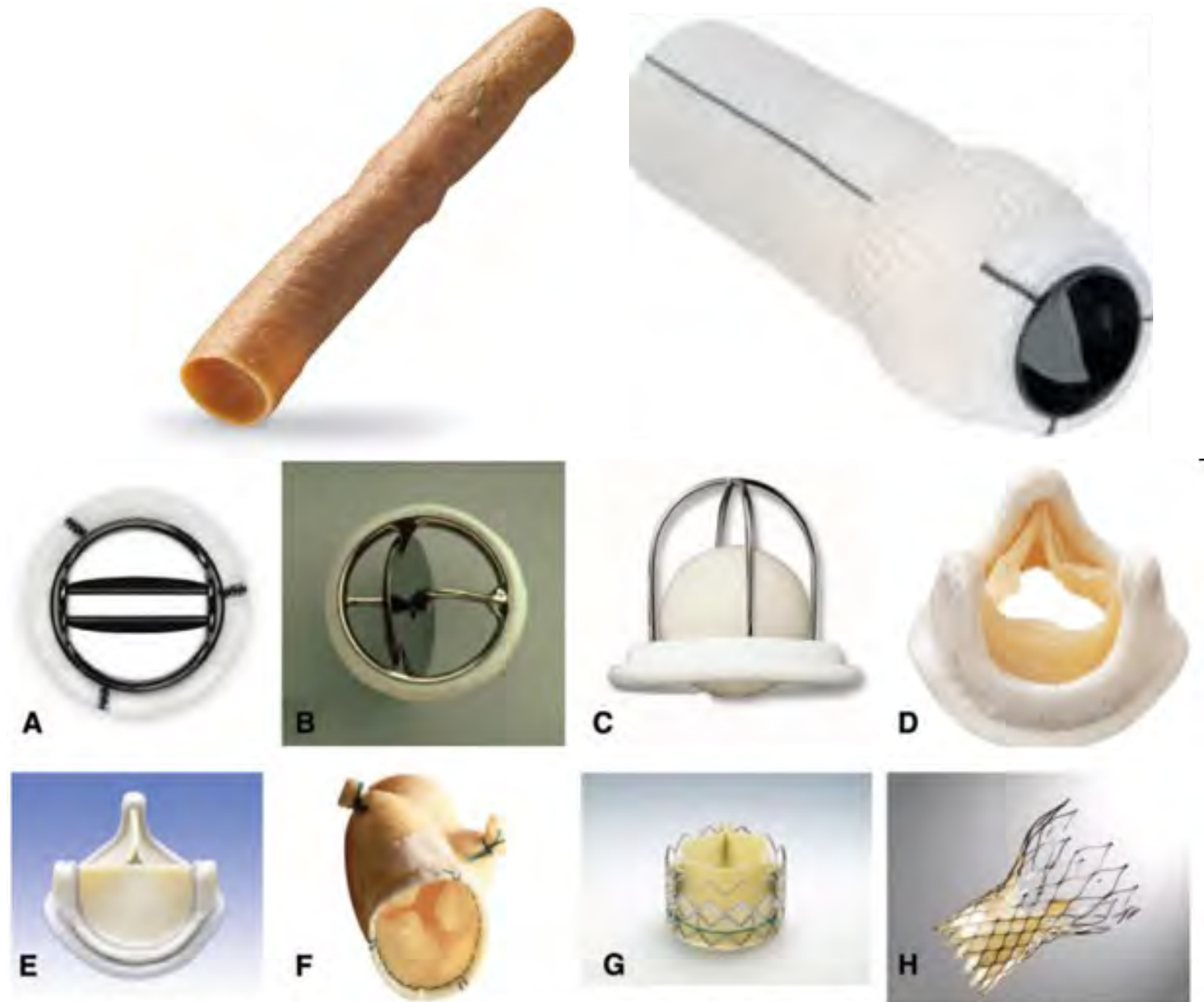
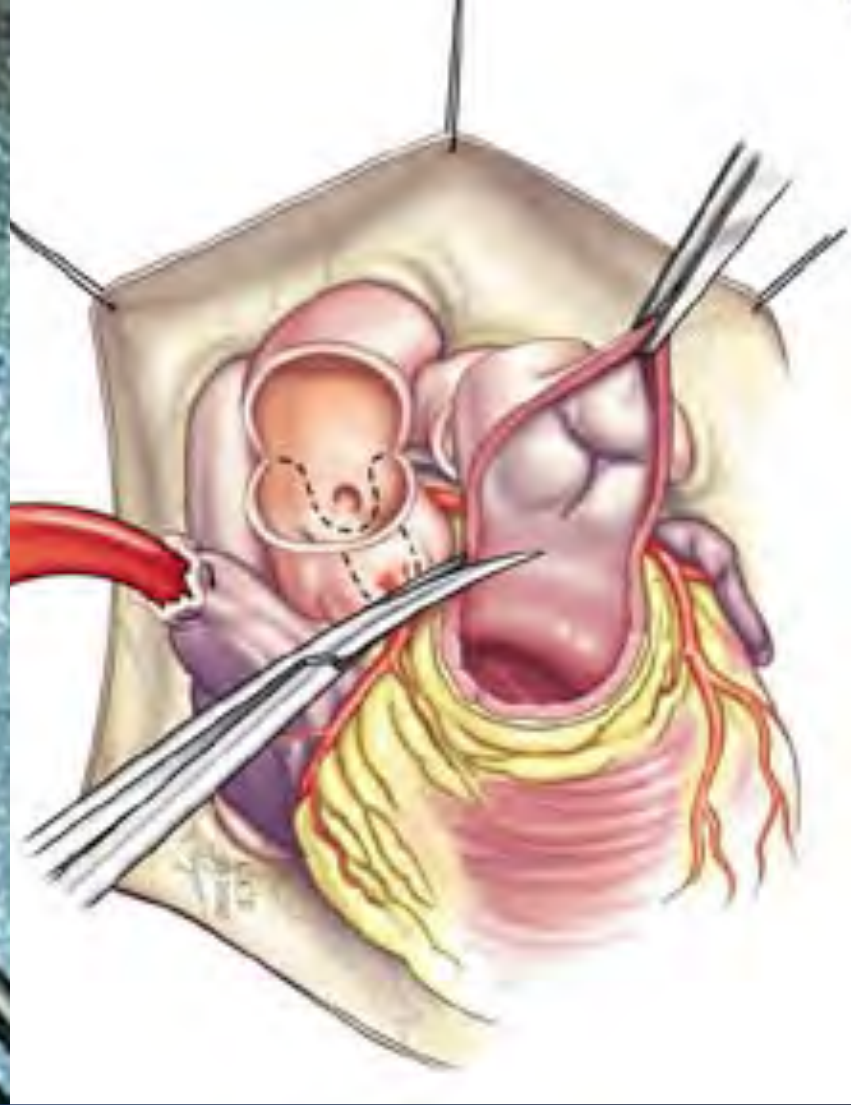


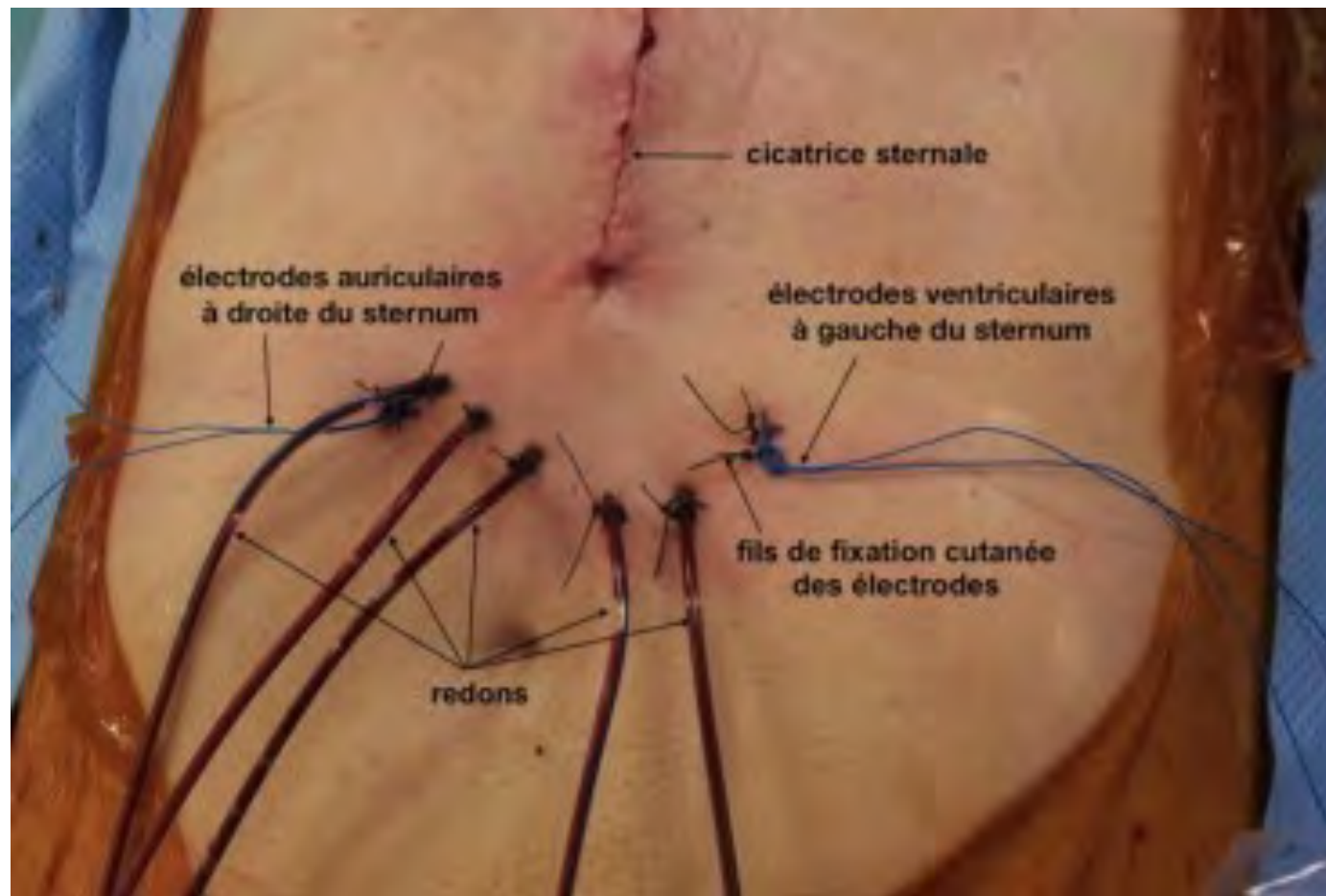
Figure 1. Different types of prosthetic valves. A, Bileaflet mechanical valve (St Jude); B, monoleaflet mechanical valve (Medtronic Hall); C, caged ball valve (Starr-Edwards); D, stented porcine bioprosthesis (Medtronic Mosaic); E, stented pericardial bioprosthesis (Carpentier-Edwards Magna); F, stentless porcine bioprosthesis (Medtronic Freestyle); G, percutaneous bioprosthesis expanded over a balloon (Edwards Sapien); H, self-expandable percutaneous bioprosthesis (CoreValve).



Valves

Matériel intra-thoracique

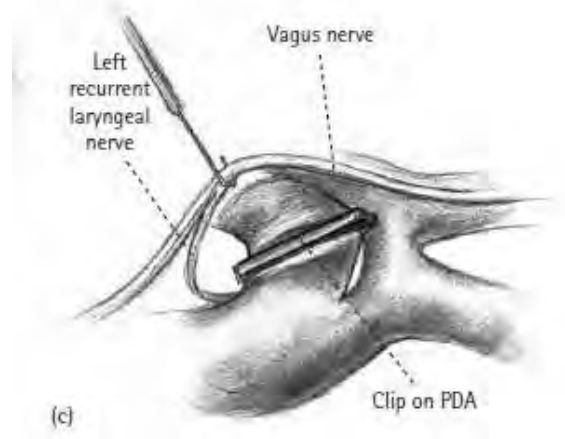
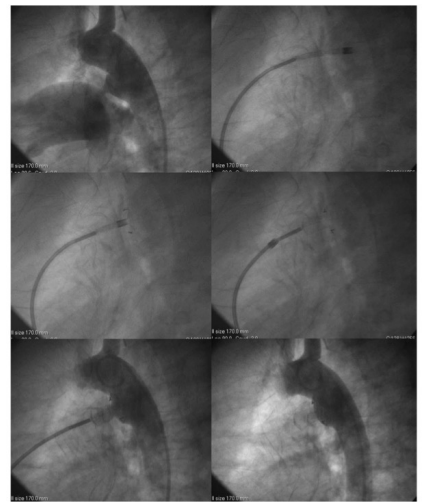
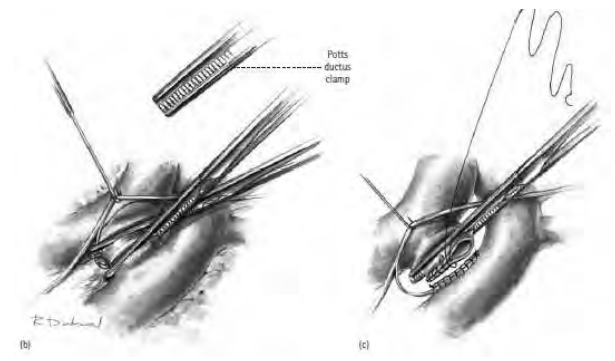
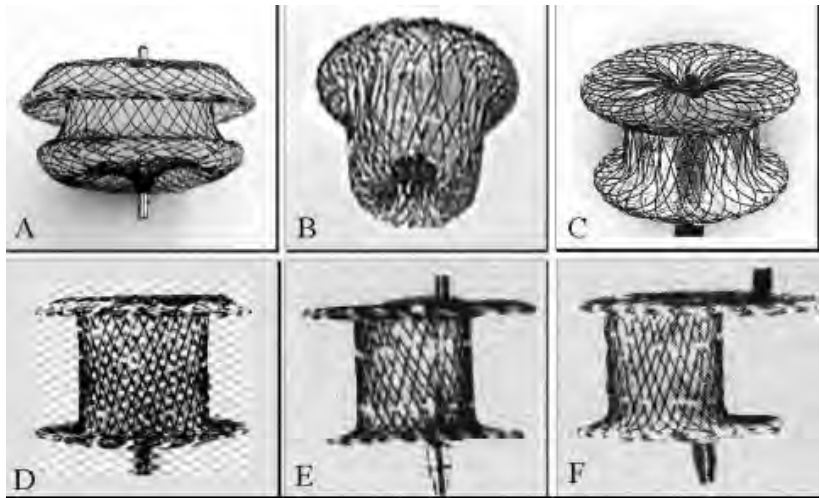
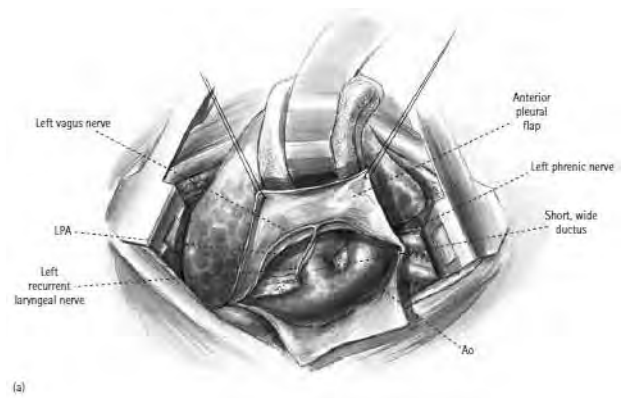
- Electrodes de stimulation
 - Auriculaires, ventriculaires
- Drainage
 - Redon ou drain pleural, médiastinal
- KT transthoracique
 - KT OG, KT AP





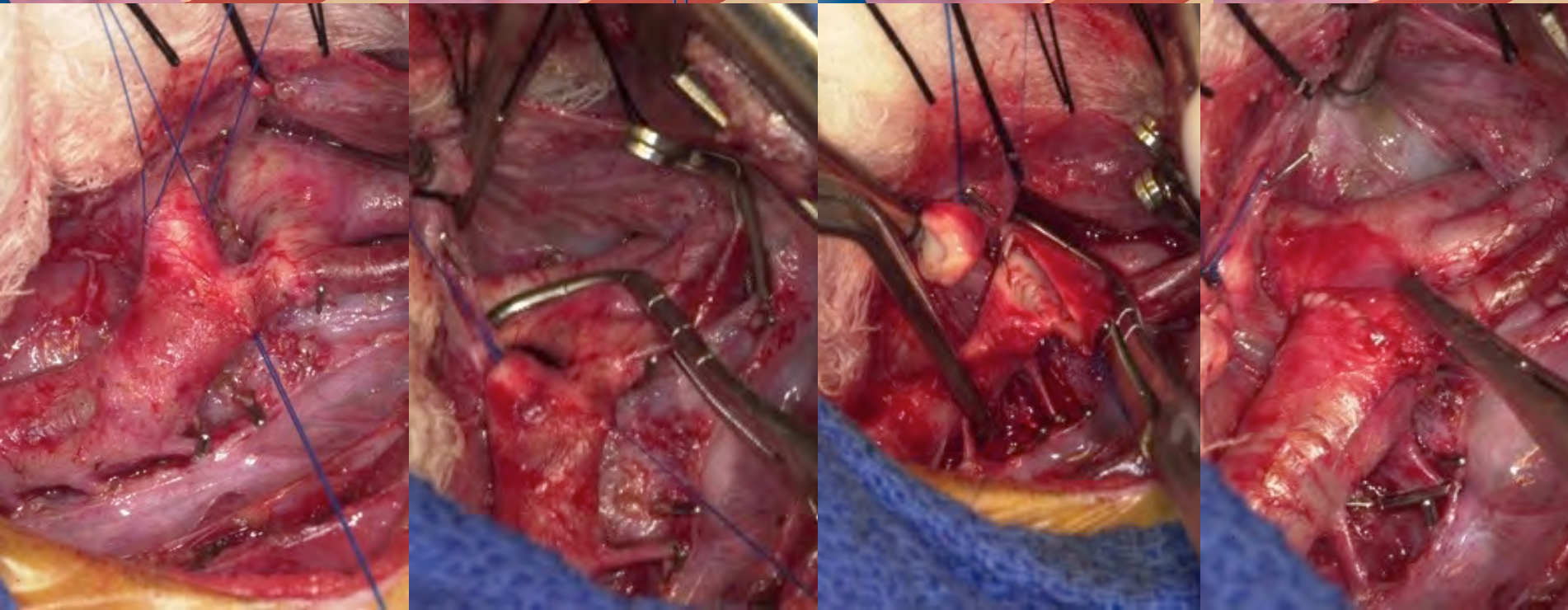
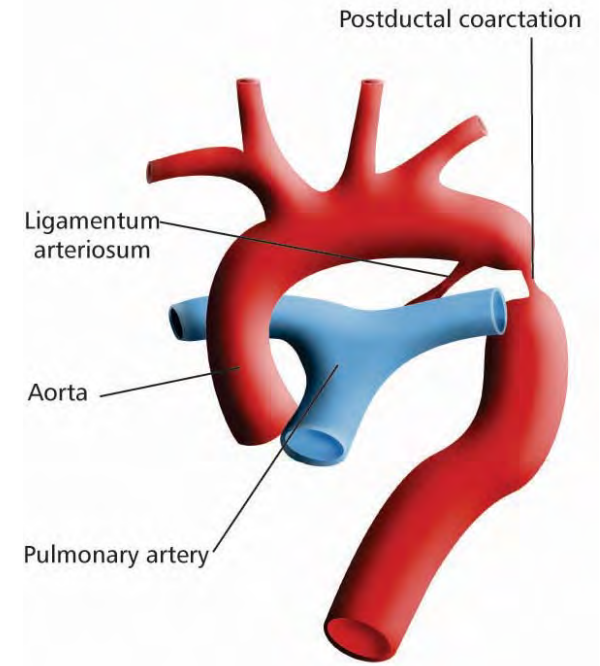
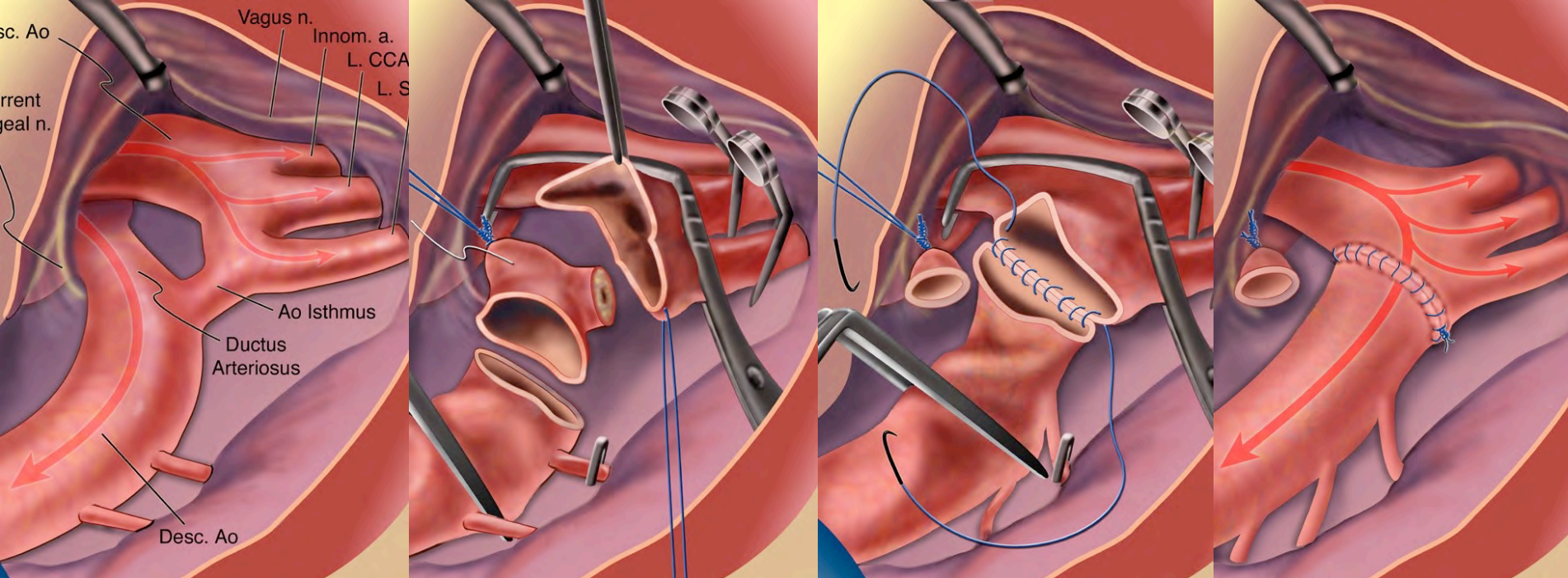
Techniques Chirurgicales

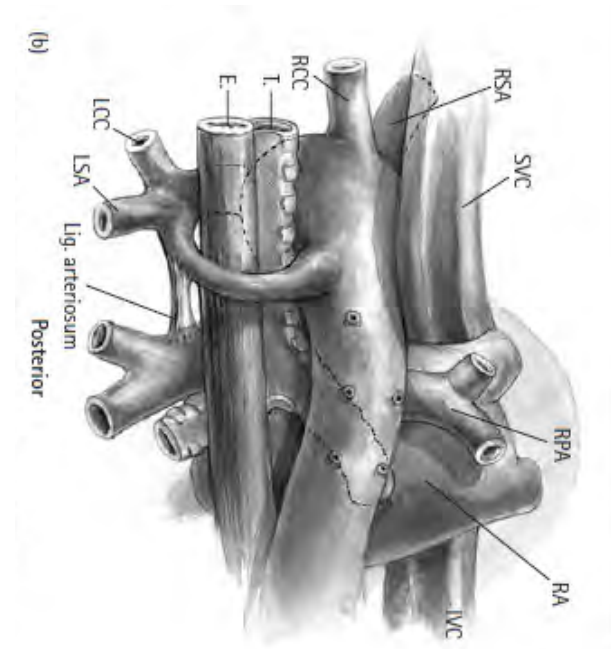




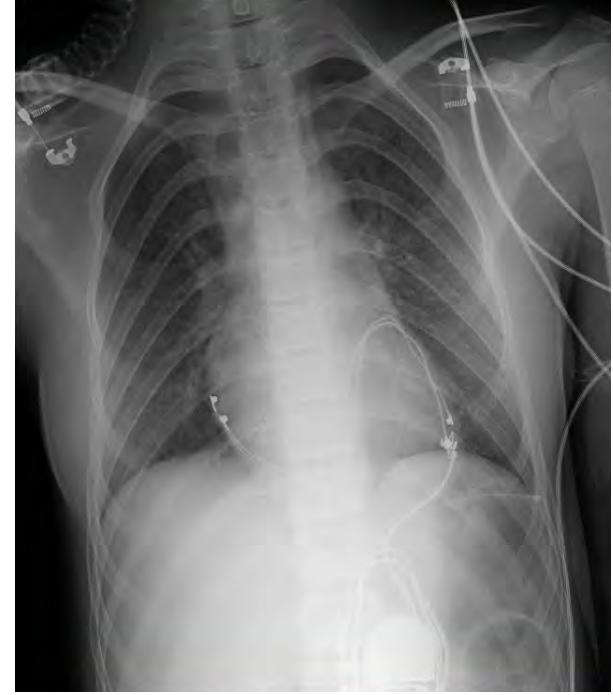
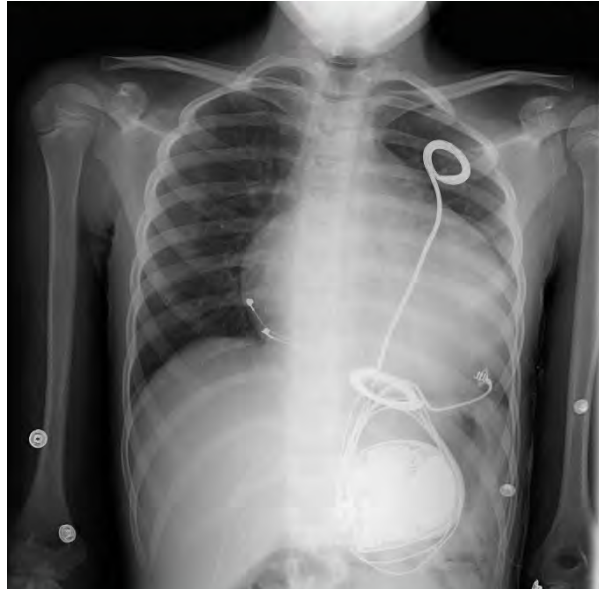
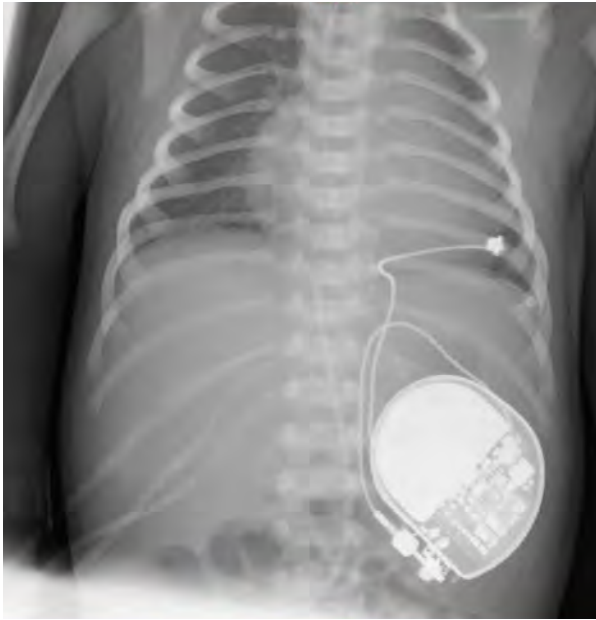
Canal Artériel

Coarctation



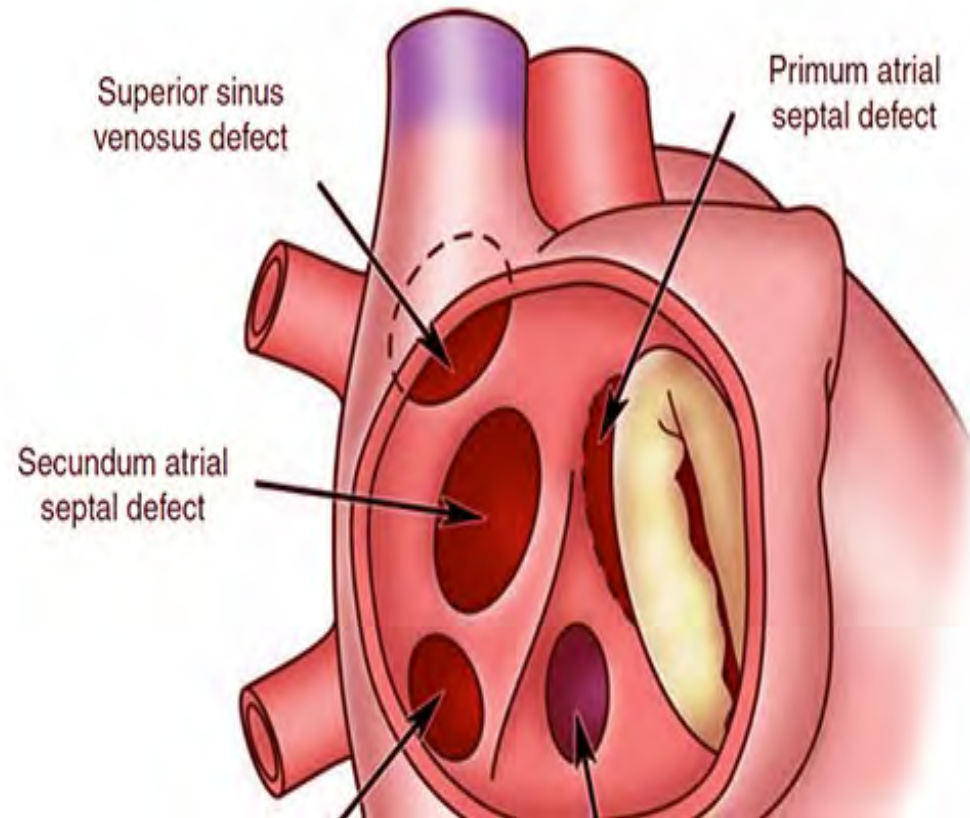


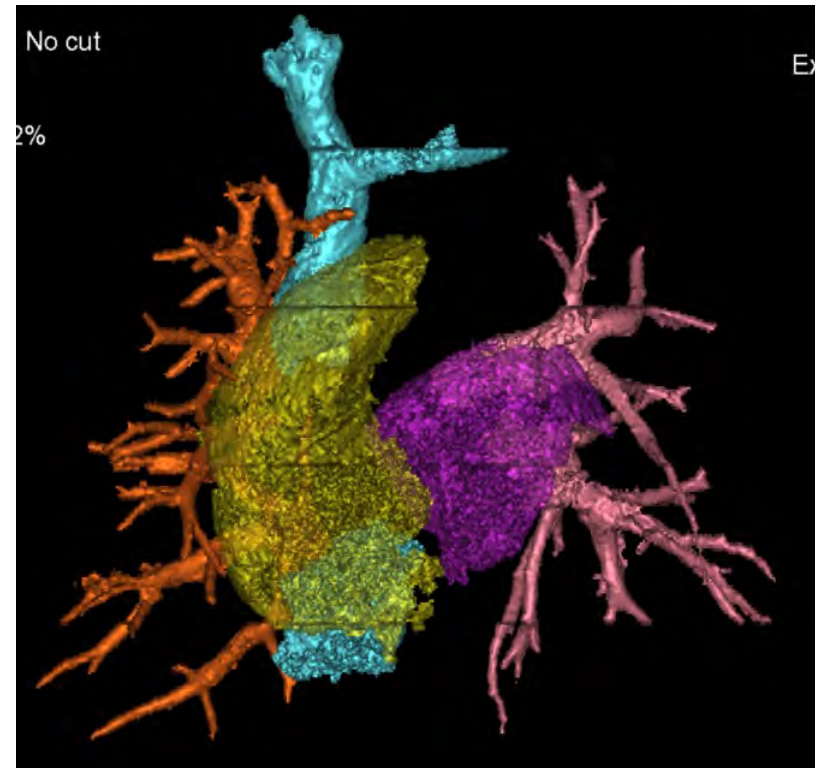
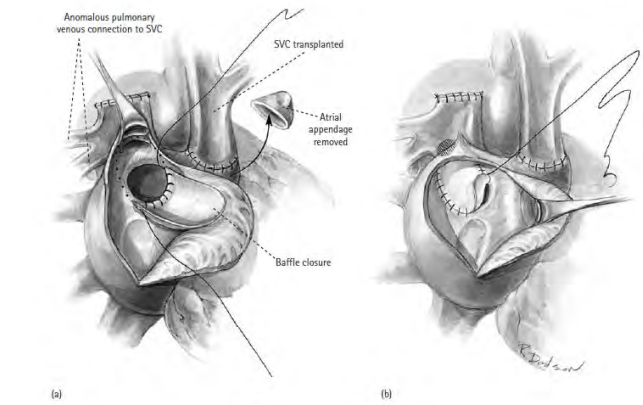
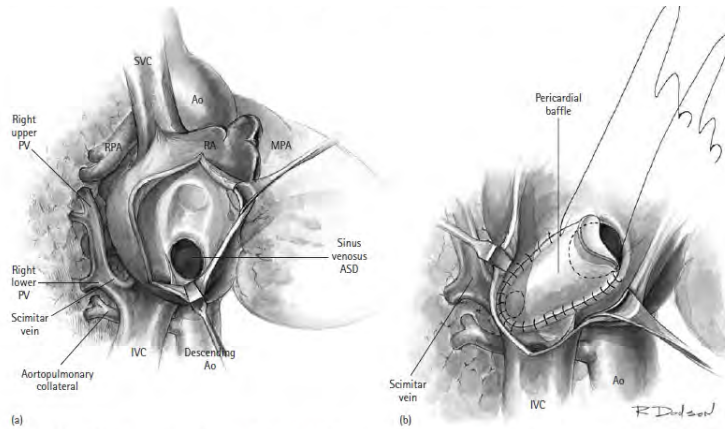
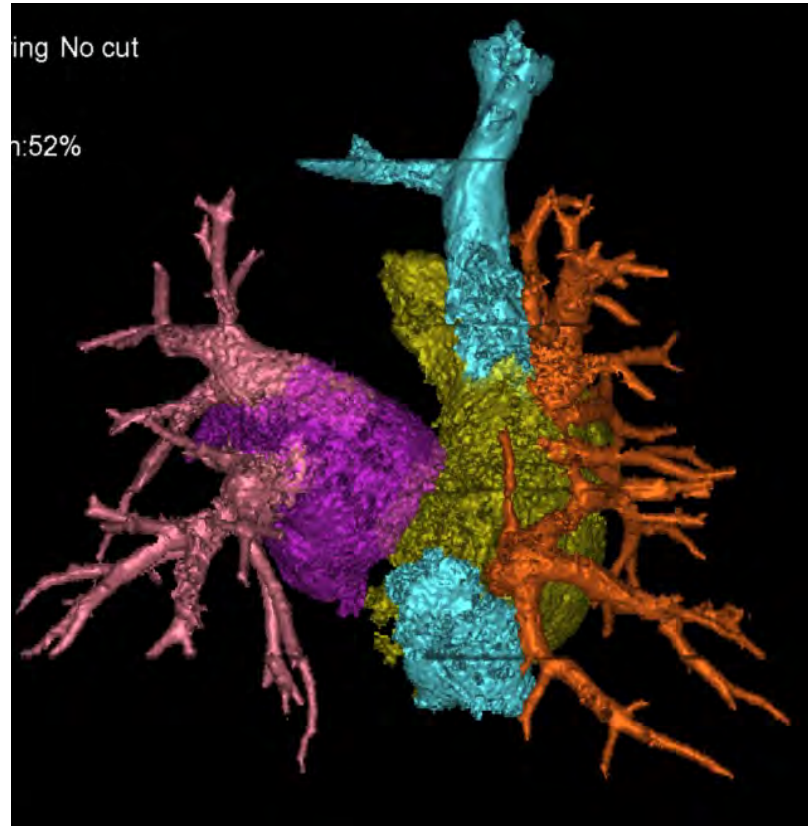
Anomalies des arcs aortiques



Rythmologie pédiatrique

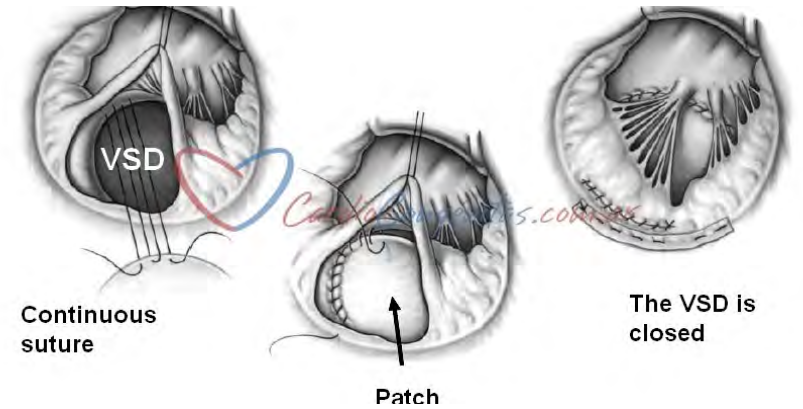
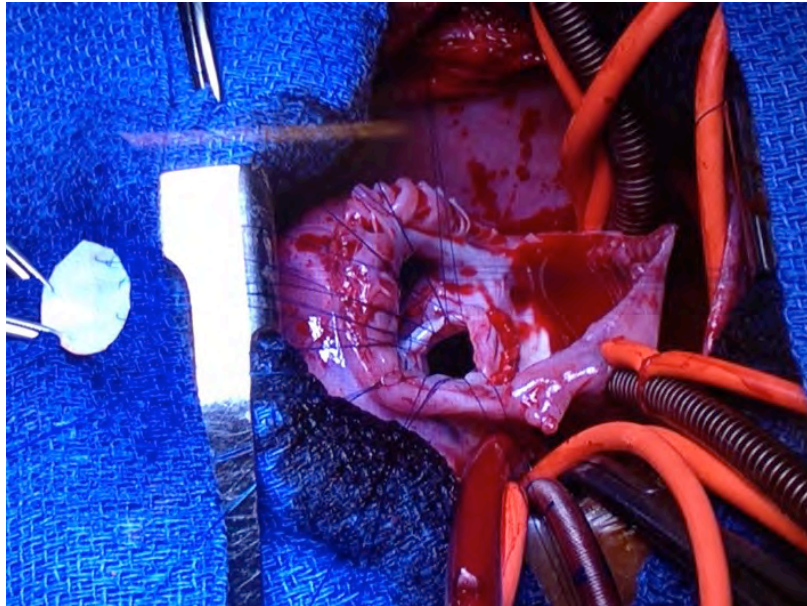
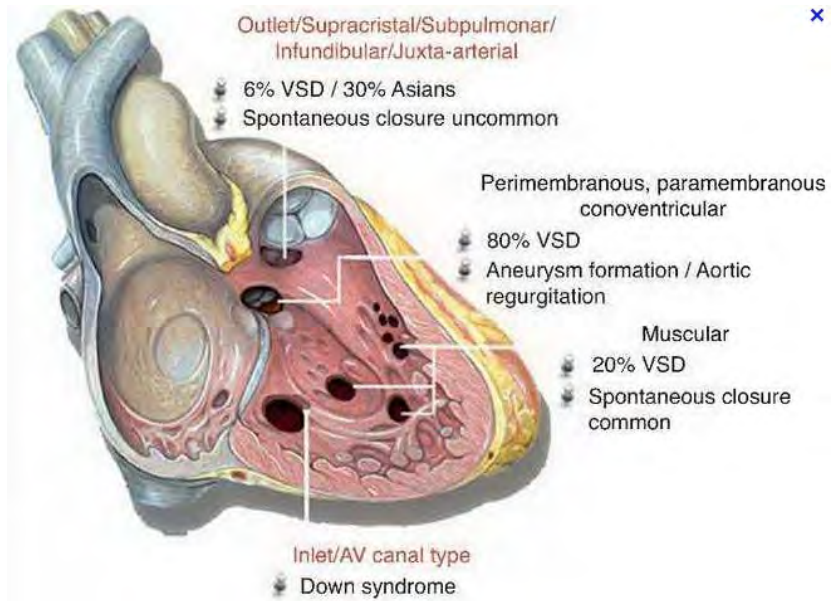
CIA



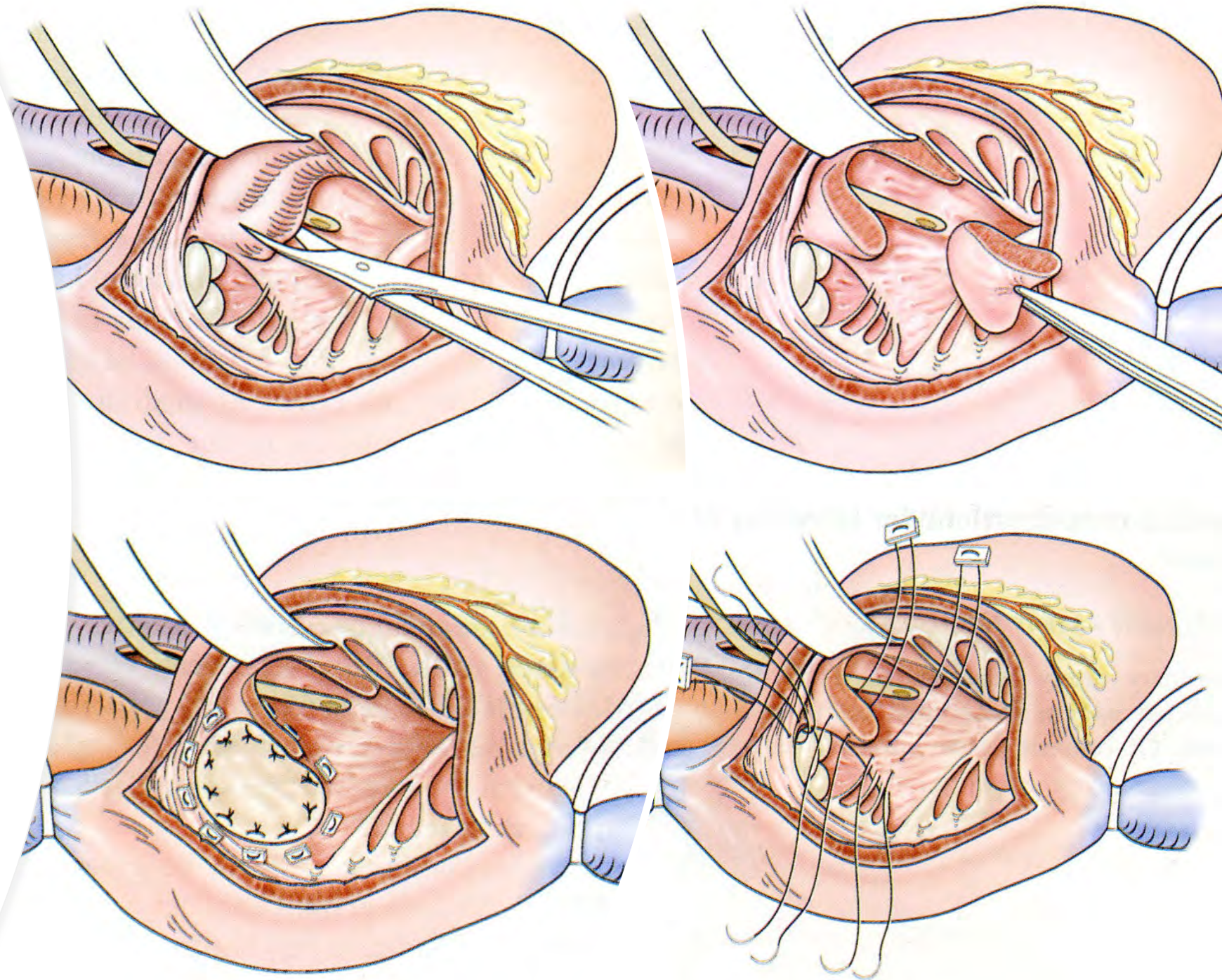


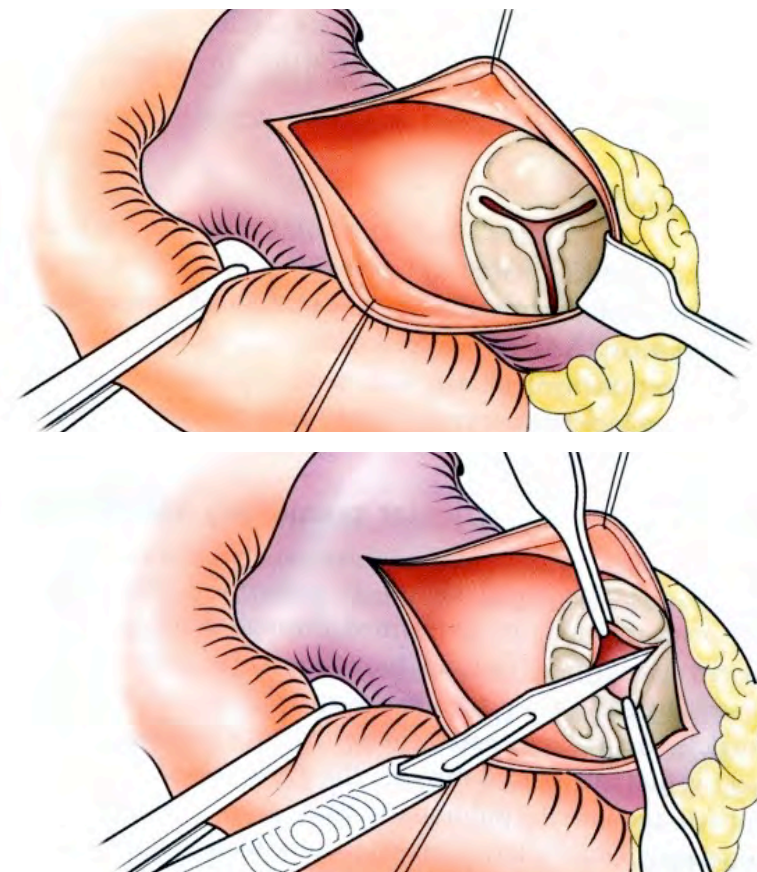
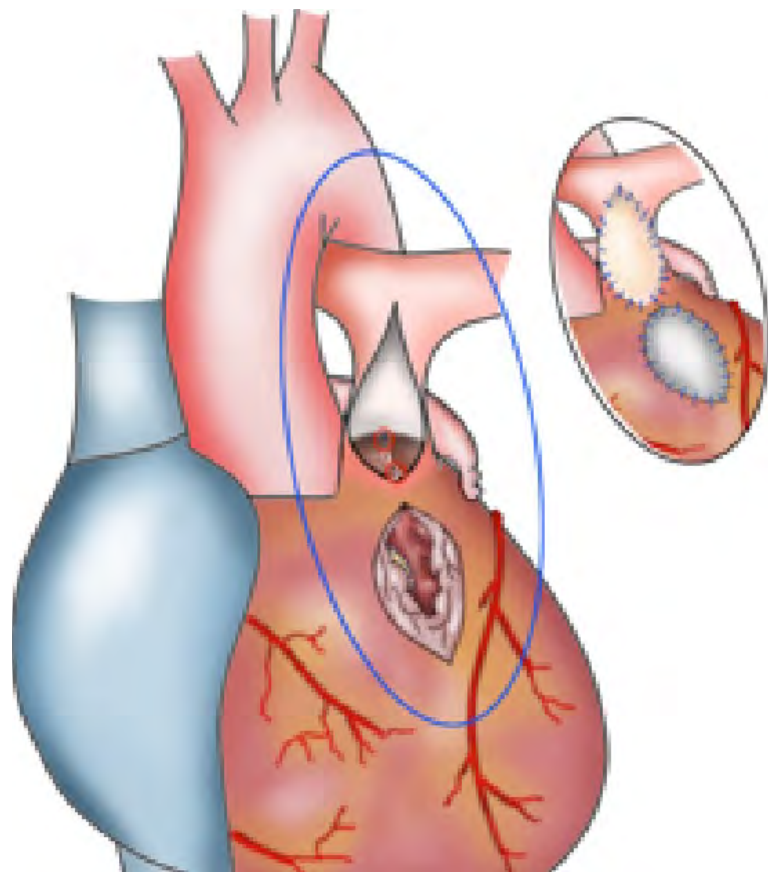
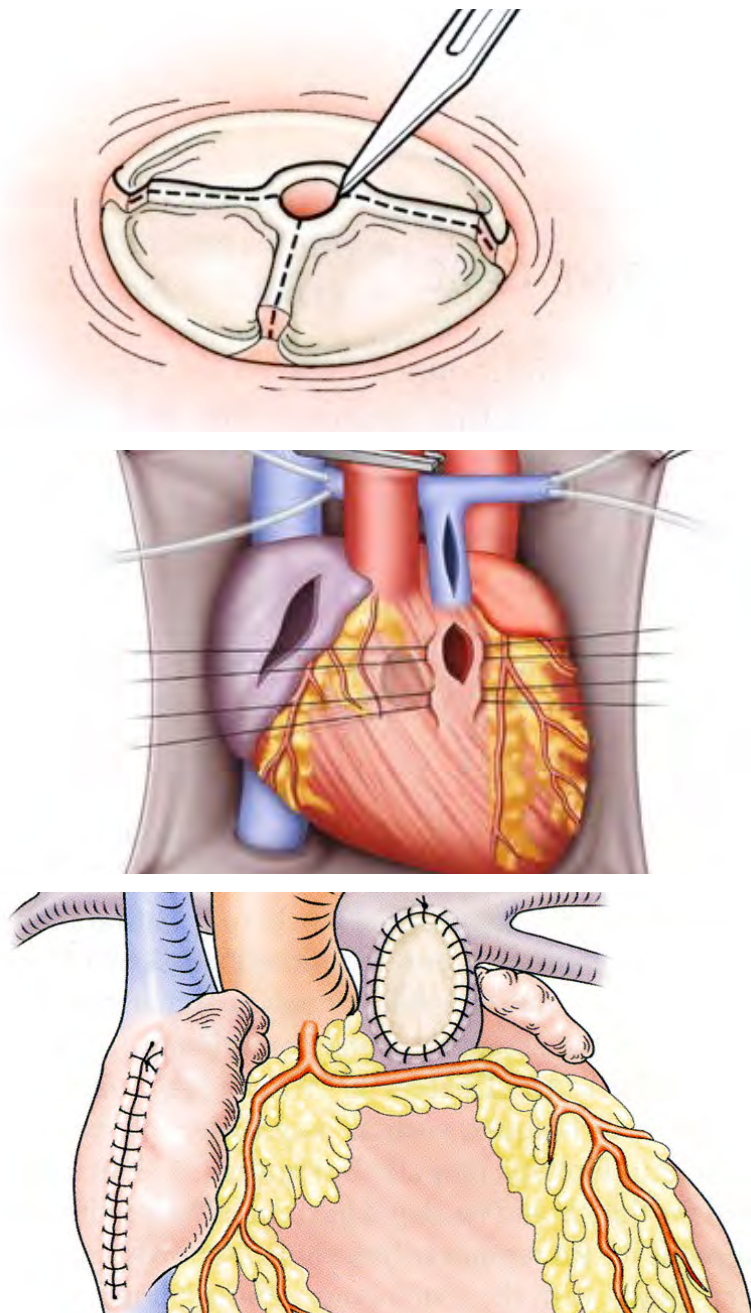
CIA et RVPAP

CIV



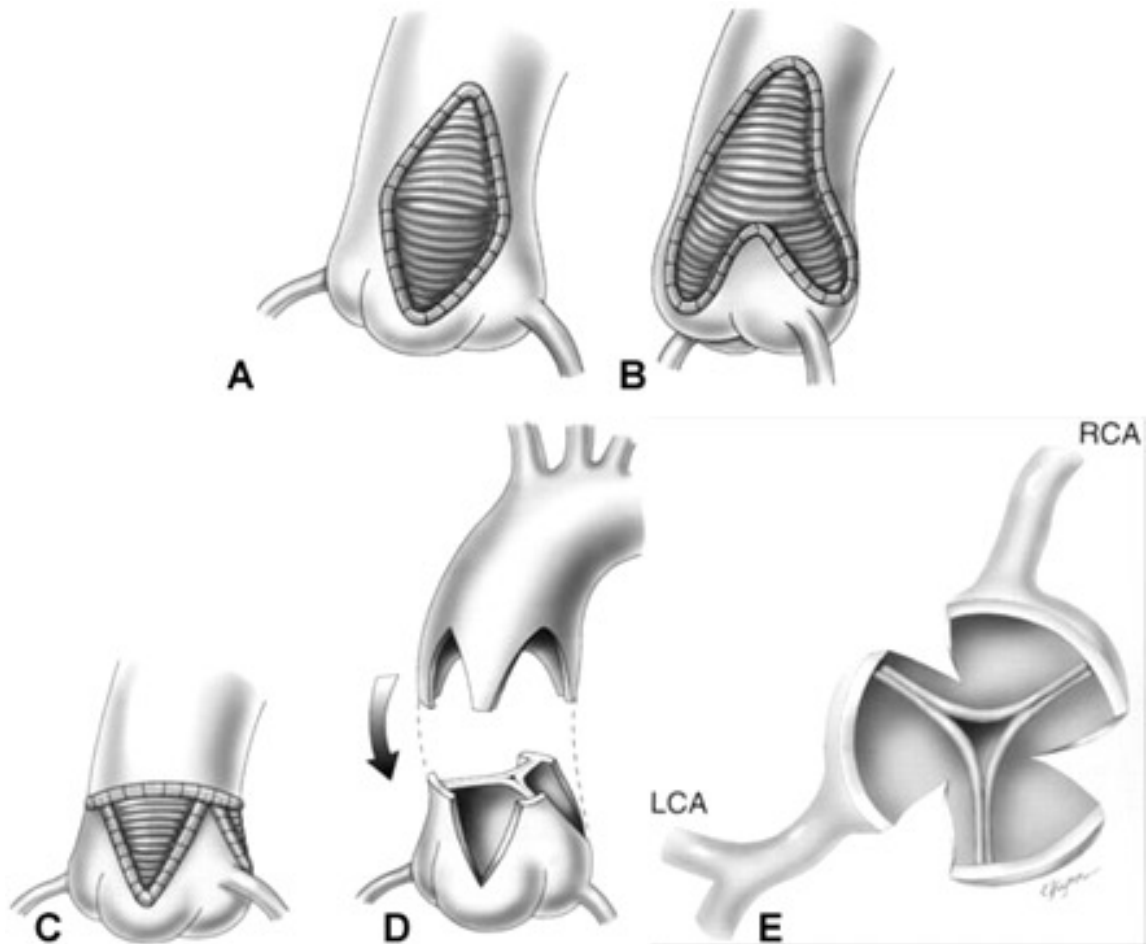
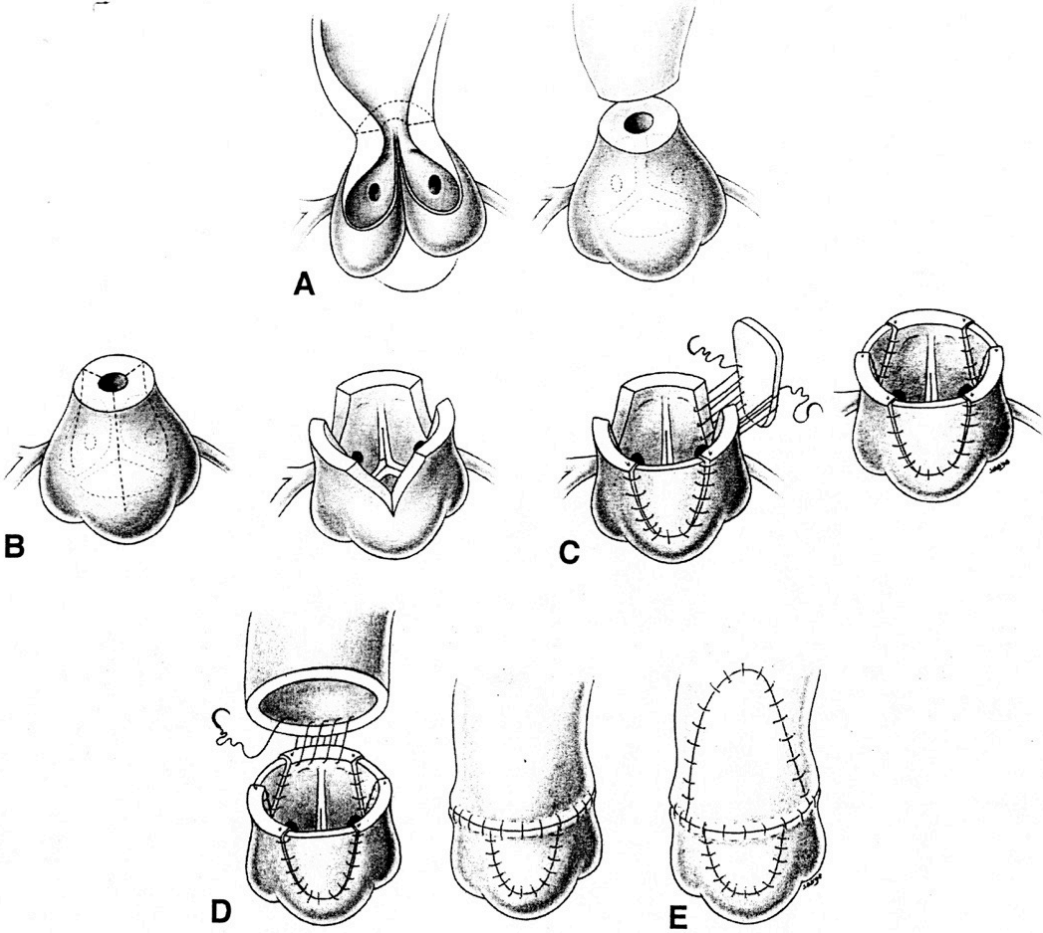
Tétralogie de Fallot



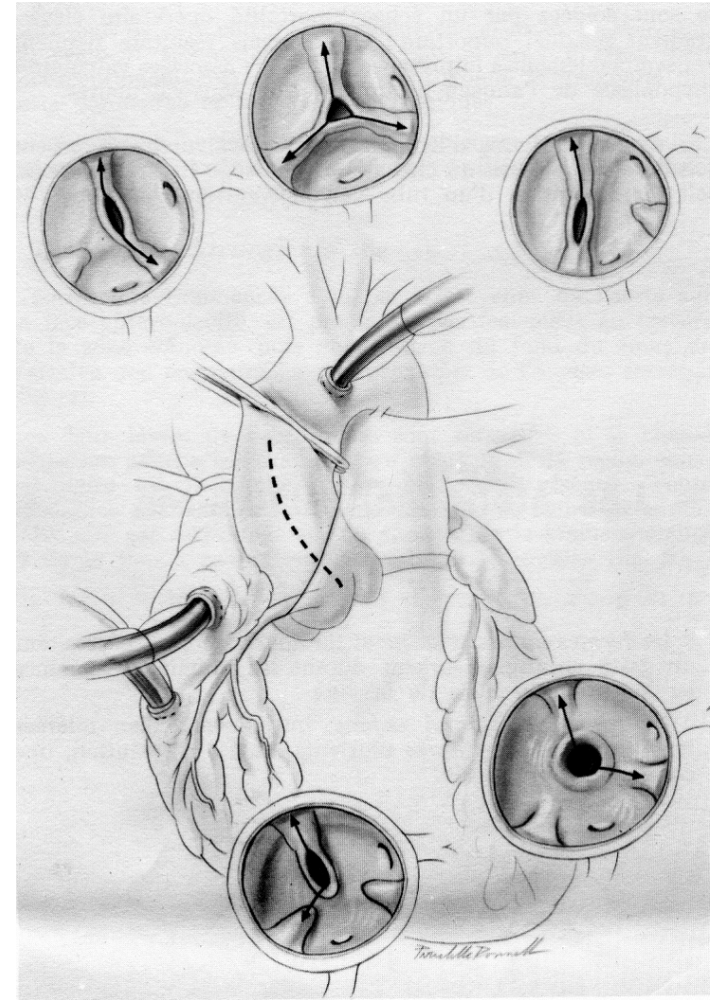
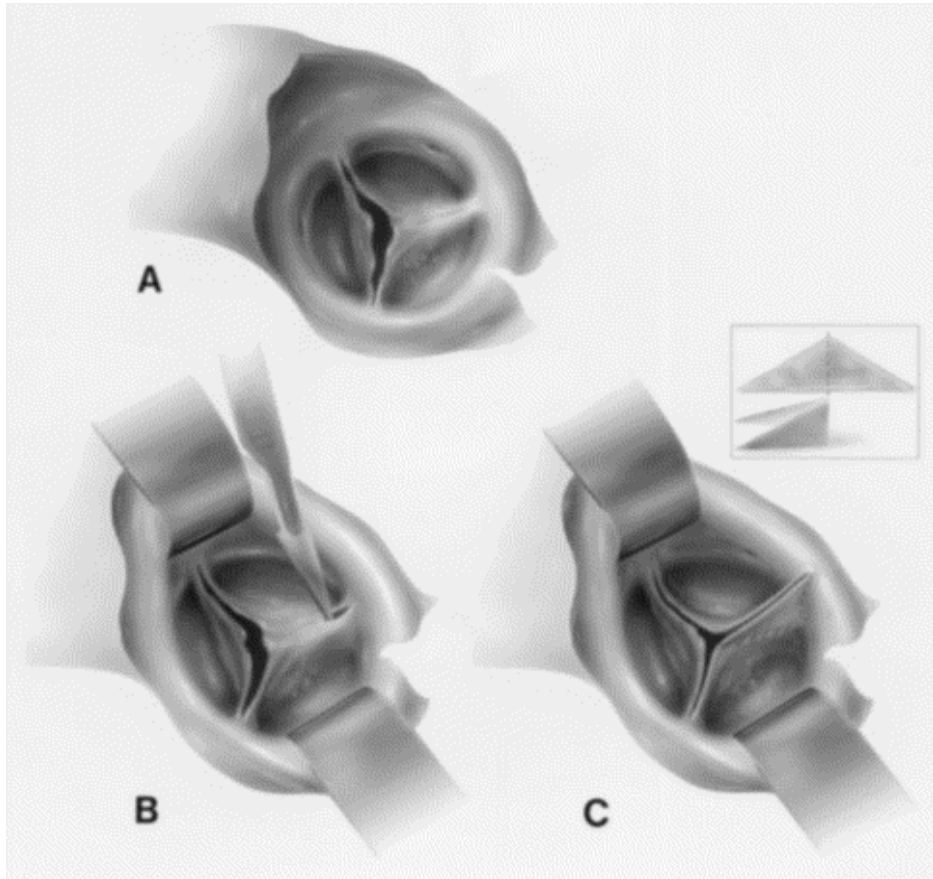


Tétralogie de Fallot – Voie droite

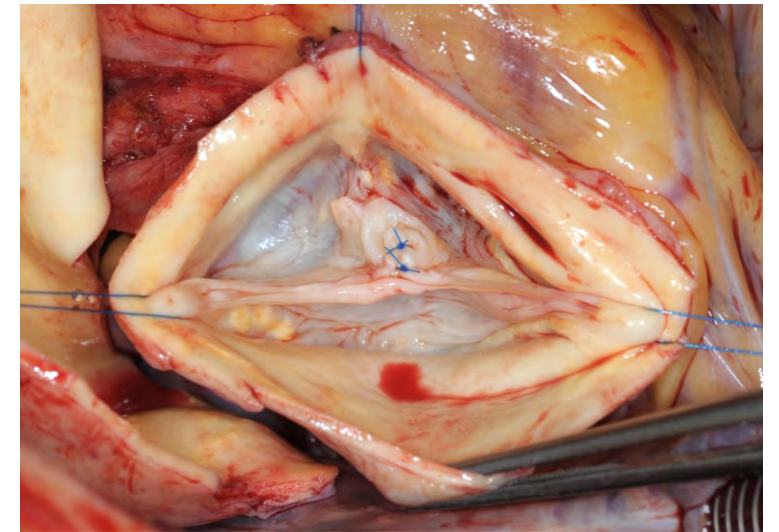
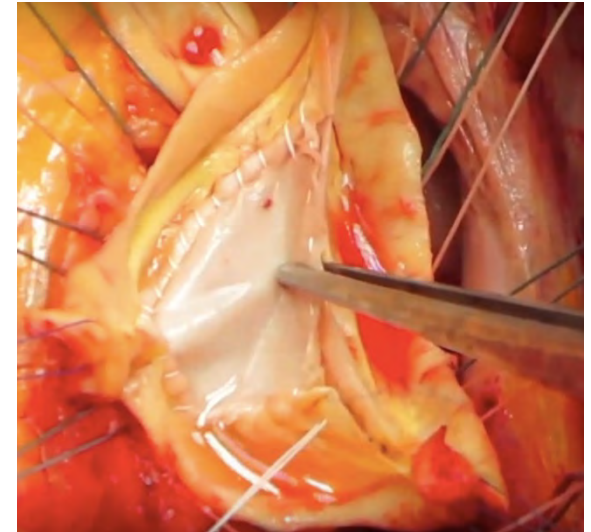
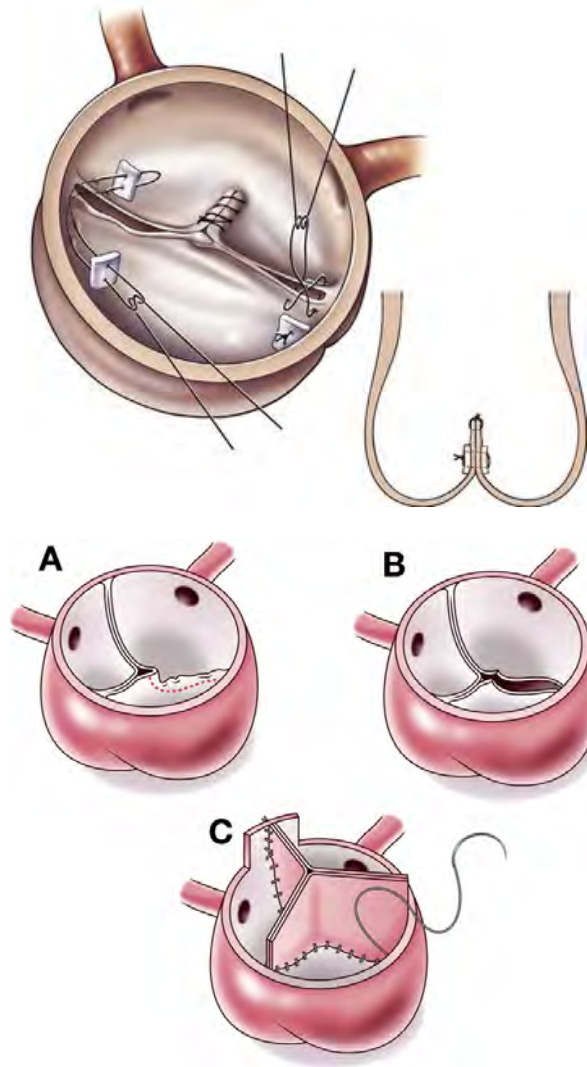
Sténoses Supra-Valvulaires

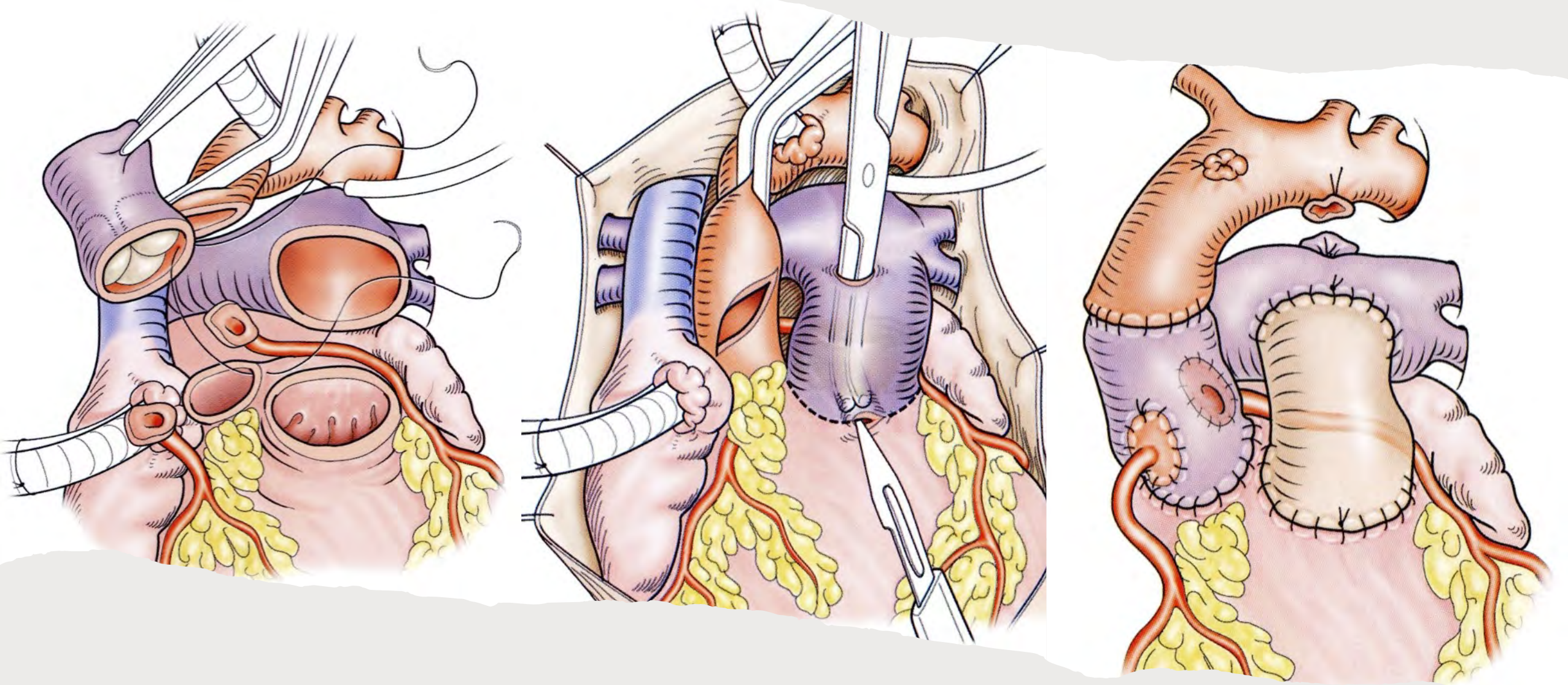


Sténoses Valvulaires Aortiques - Commissurotomie



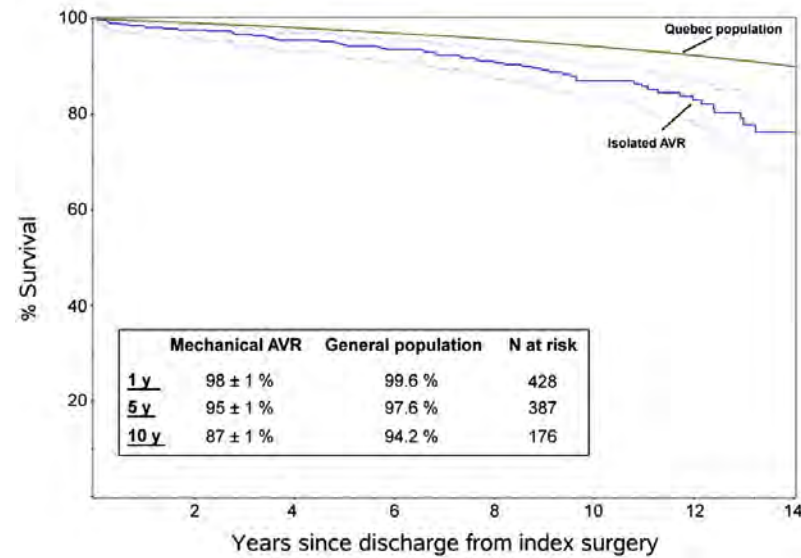
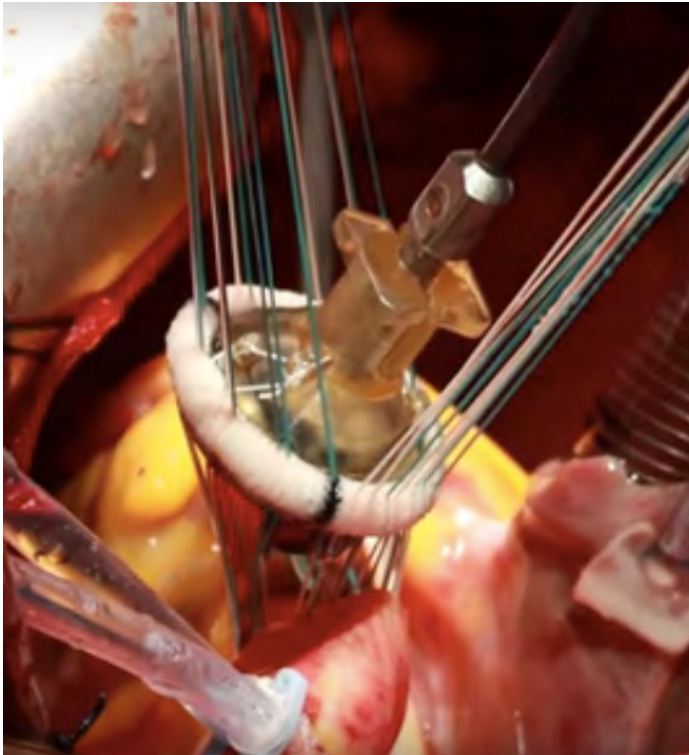
Sténoses Valvulaires Aortiques - Plastie





Sténoses Valvulaires Aortiques - Ross

Sténoses Valvulaires Aortiques - RVAo



The Journal of Thoracic and Cardiovascular
Surgery

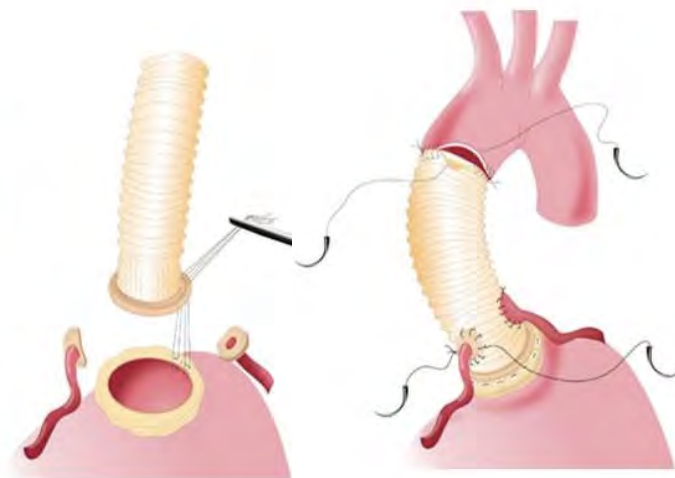
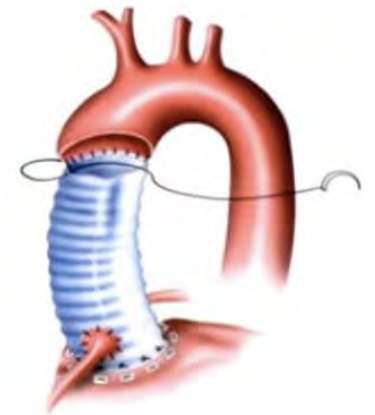
Volume 148, Issue 4, October 2014, Pages 1341-1346.e1

Acquired cardiovascular disease

Long-term outcomes after elective isolated
mechanical aortic valve replacement in
young adults

Ismail Bouhout MSc^a, Louis-Mathieu Stevens MD, PhD^b, Amine Mazine MSc^a, Nancy Poirier MD^a, Raymond Cartier MD^a, Philippe Demers MD^a, Ismail El-Hamamsy MD, PhD^a

Insuffisance Valvulaire Aortique



Remodeling & Annuloplasty



[TAV: Article 1 of series⁹](#)

[BAV: Current Article - 2 of series](#)

Asc Aorta Replacement & Annuloplasty



[TAV: Article 3 of series](#)

[BAV: Article 4 of series](#)

Double Annuloplasty



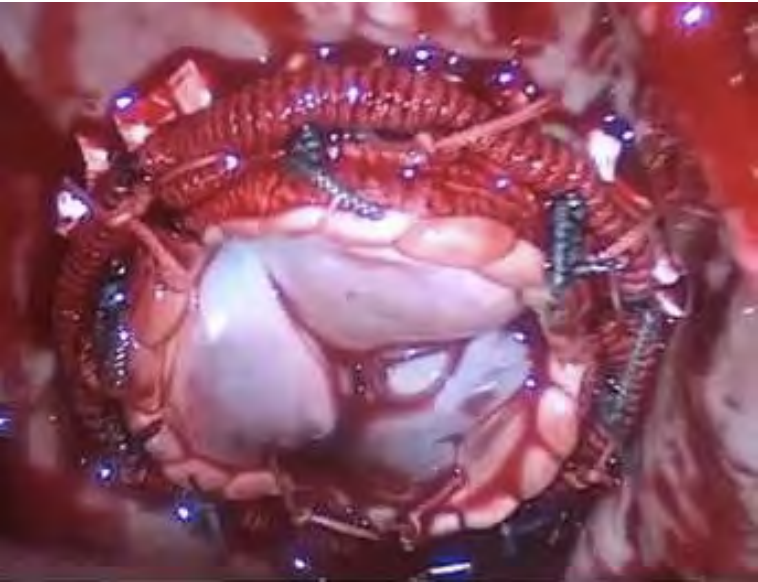
[TAV: Article 3 of series](#)

[BAV: Article 4 of series](#)

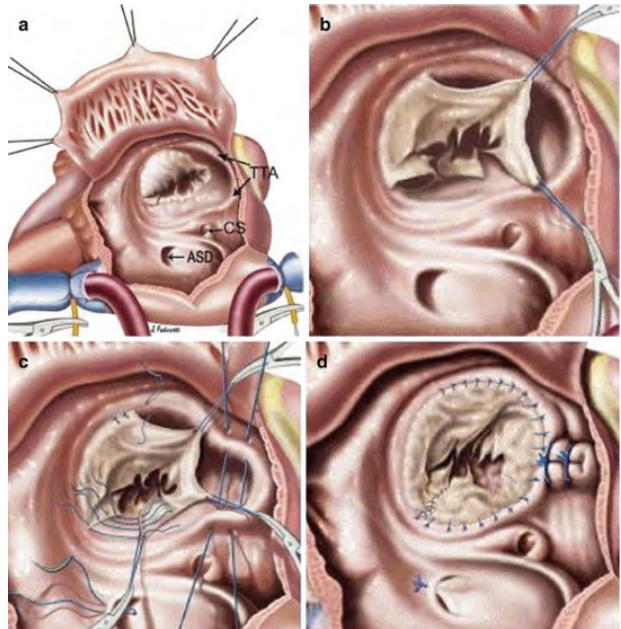
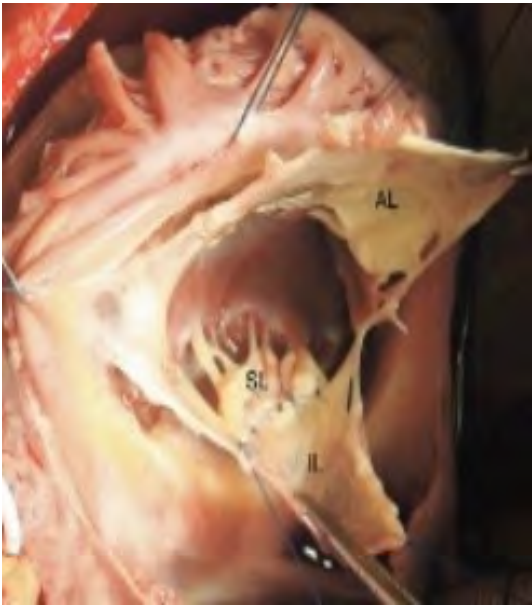
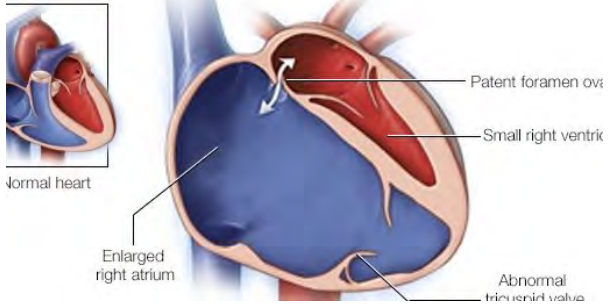
Remplacement valvulaire mécanique

- RVA mécaniques
 - Mortalité précoce 5%
 - Survie moyenne 93% à 5 ans et 10 ans
 - Survie sans réintervention 87% à 10 ans
- RVM mécaniques
 - Mortalité précoce 9%
 - Survie moyenne 82% à 5 ans, 79% à 10 ans
 - Survie sans réintervention 68% à 10 ans
- PM 11%, 3,5% endocardite, 2% thrombose de valve, 7% saignement grave, 6% embolie

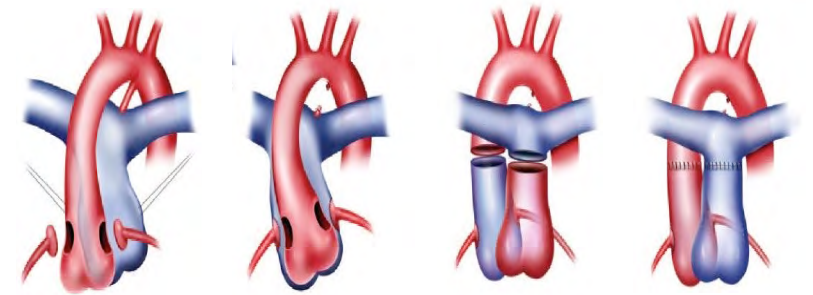
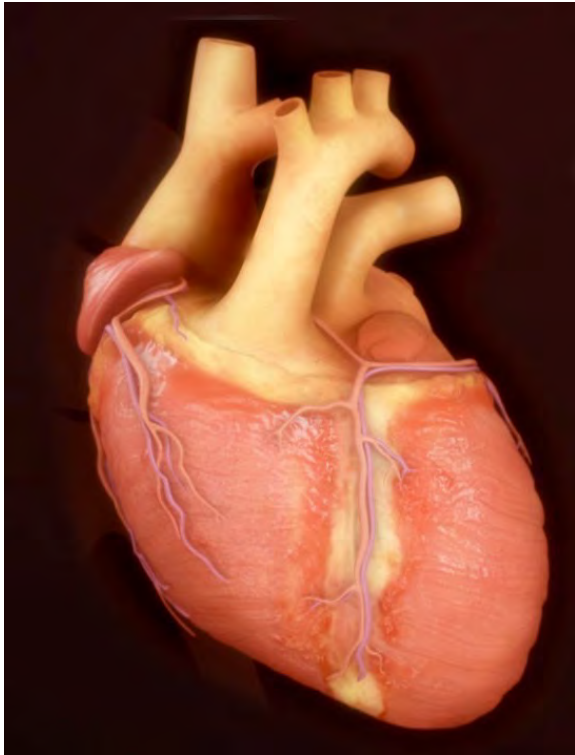
Pathologie tricuspide



EBSTEIN ANOMALY



Transposition des Gros Vaisseaux

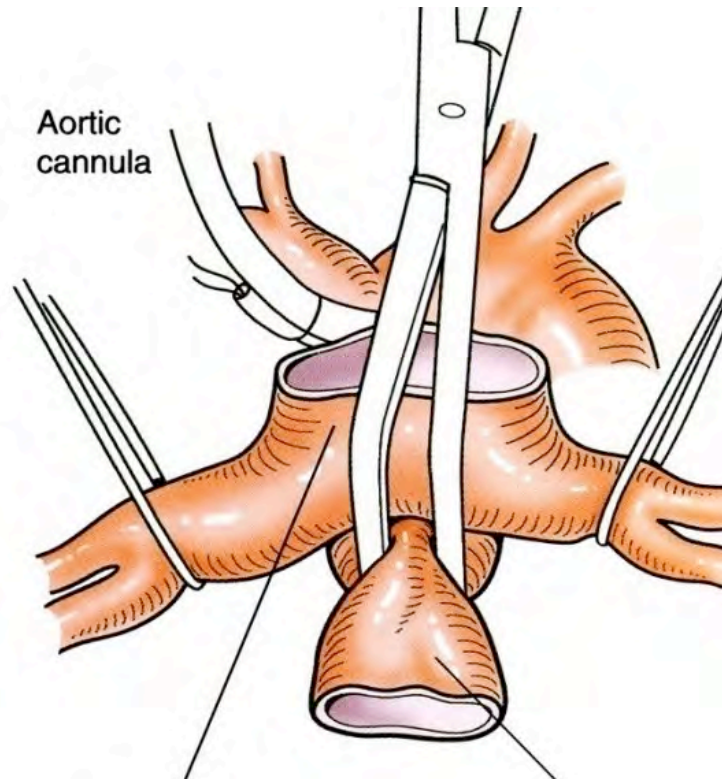


TGV – Section Aortique et Pulmonaire

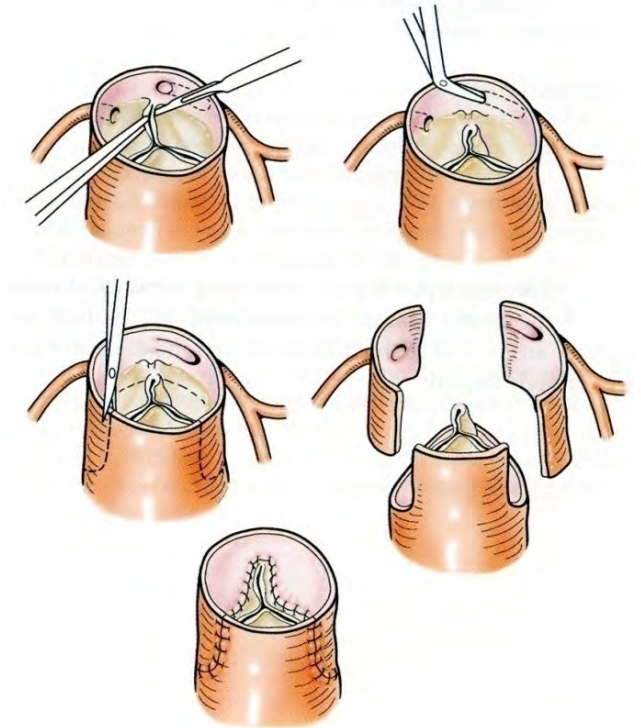
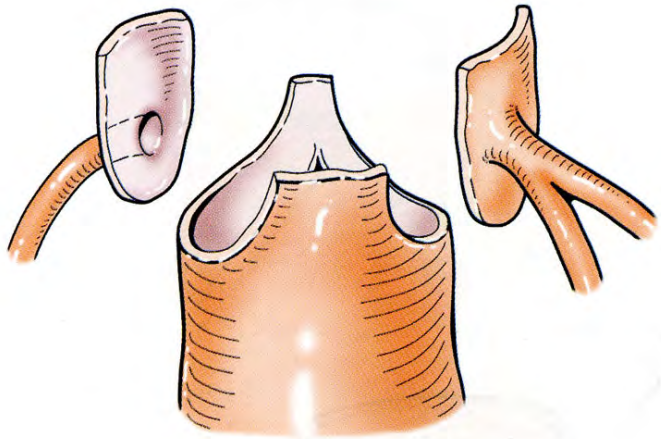
- Aortique:
 - à distance de la néo-valve pulmonaire
- Pulmonaire:
 - au dessus des commissures
 - exploration des néo-cusp aortique
 - recherche d'un malignement commissural

TGV - Manœuvre de Lecompte

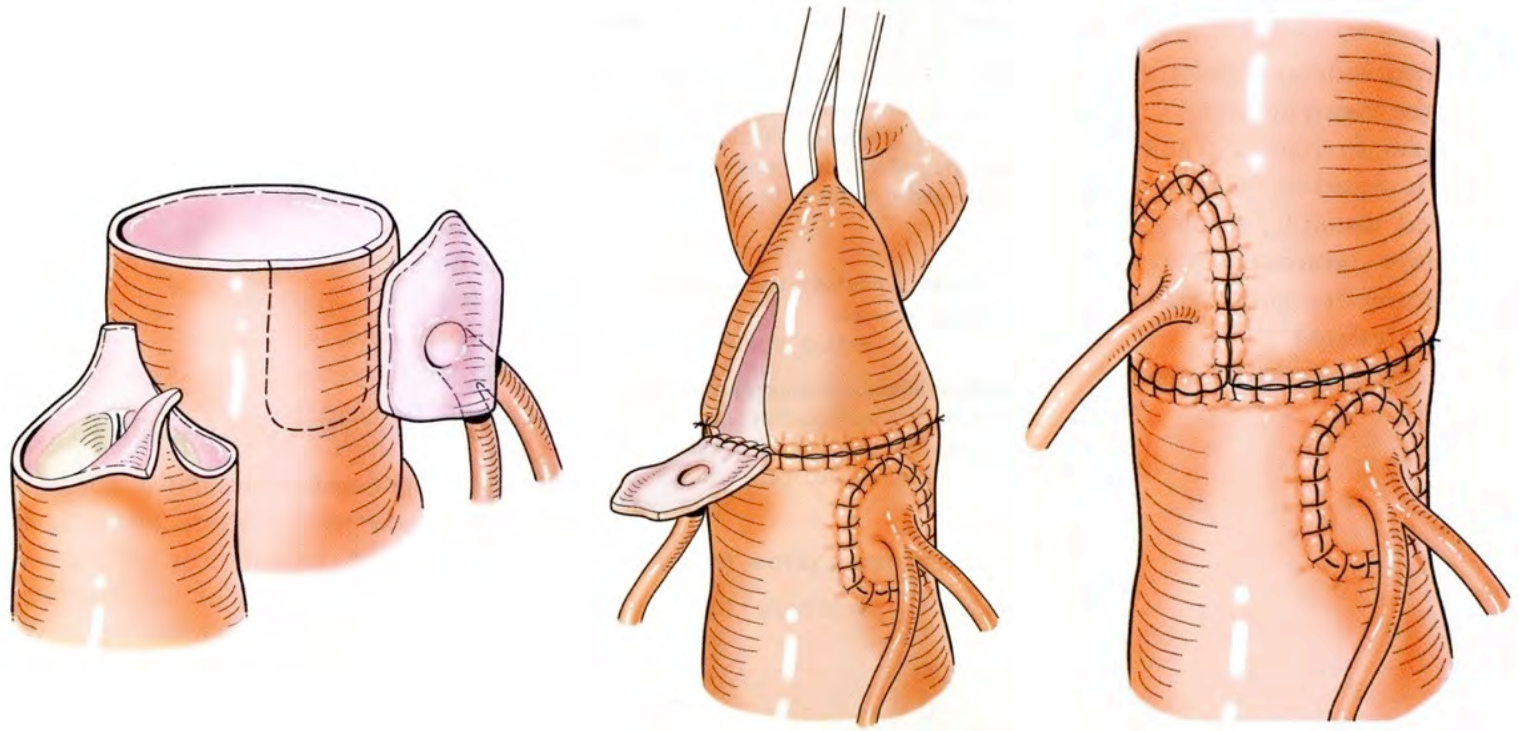
- Mobilisation extensive des AP
- Section distale de l'aorte
- Néo-aorte courte



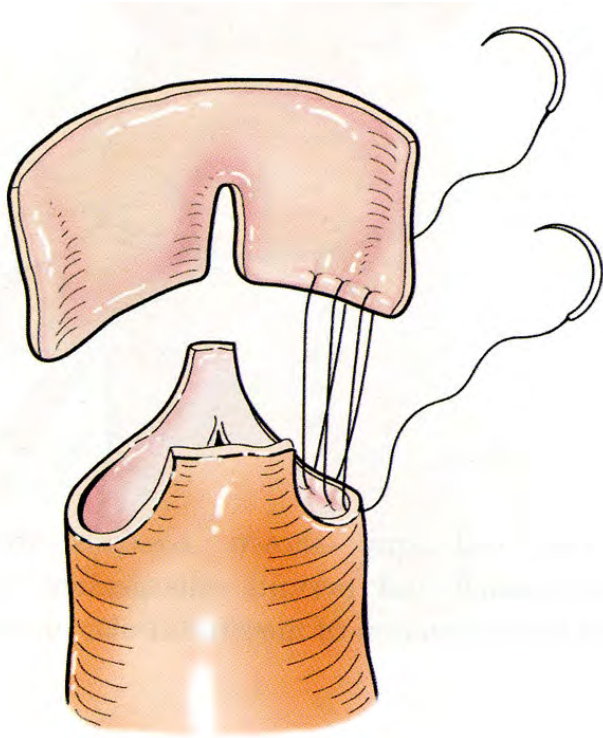
TGV - Prélèvement des coronaires



TGV – Réimplantation des Coronaires



TGV – Reconstruction Pulmonaire





Spécificités des GUCH



Chirurgie des cardiopathies congénitales chez l'adulte

- GUCH : Grown up Congenital Heart Disease
-> ACHD : Adult Congenital Heart Disease
- Cardiopathie vieillie découverte tardivement
- Redux, tridux, quadridux... multiopéré



Due to medical, surgical, and technological evolutions,
>90% of individuals with CHD who are born, now survive into adulthood

Chirurgie des cardiopathies congénitales chez l'adulte



ESC

European Society
of Cardiology

European Heart Journal (2021) **42**, 563–645

doi:10.1093/eurheartj/ehaa554

ESC GUIDELINES

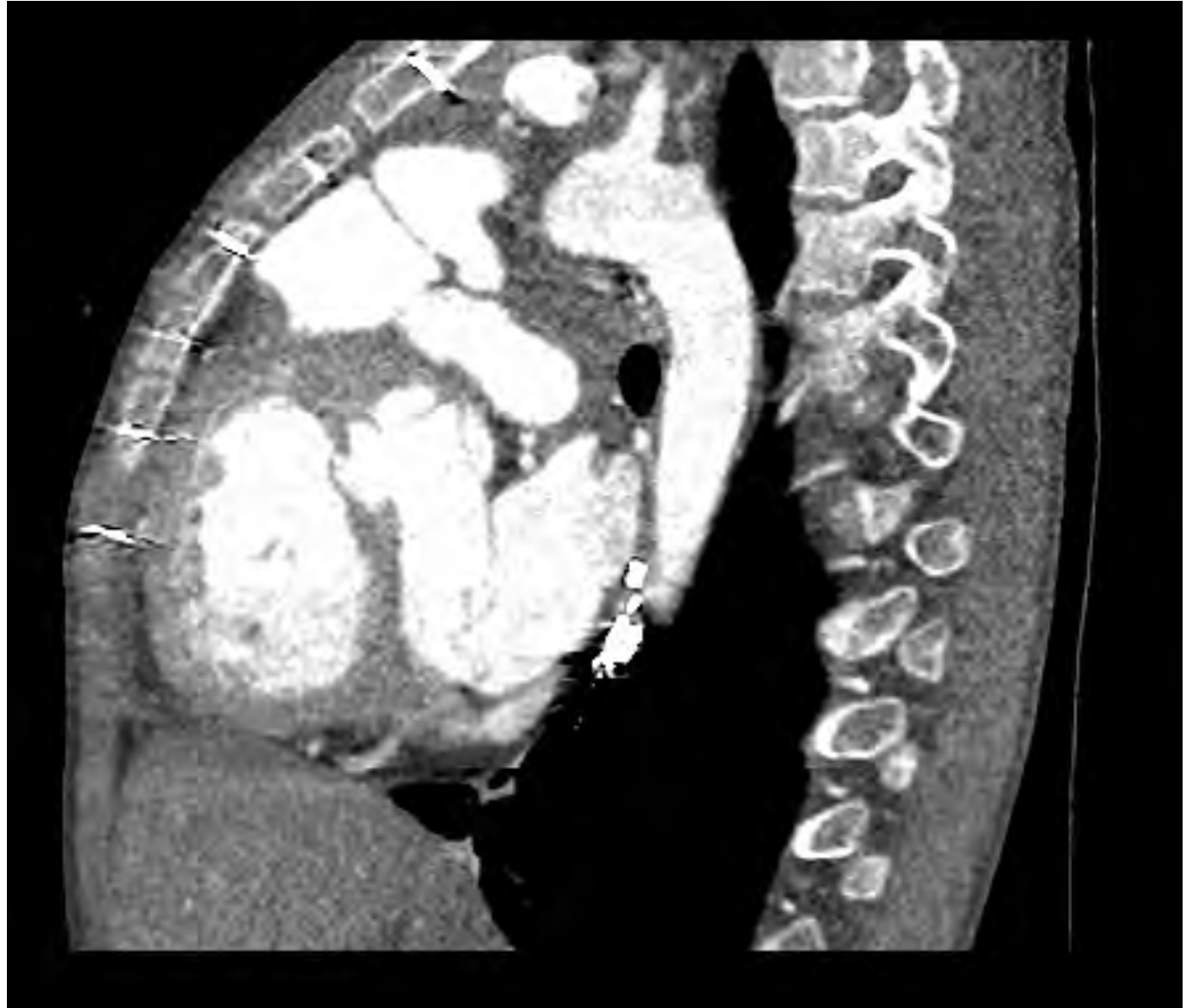
2020 ESC Guidelines for the management of adult congenital heart disease

**The Task Force for the management of adult congenital heart
disease of the European Society of Cardiology (ESC)**

**Endorsed by: Association for European Paediatric and Congenital Cardiology
(AEPC), International Society for Adult Congenital Heart Disease (ISACHD)**

Chirurgie des cardiopathies congénitales chez l'adulte

- Principalement chirurgie de la voie droite
 - Fallot, APSO
- Evaluation imagerie ++
 - Proximité des structures cardiaques avec sternum : TDM !
 - Accès vasculaire : Echo doppler !
 - Shunt persistant : ETO test aux bulles !



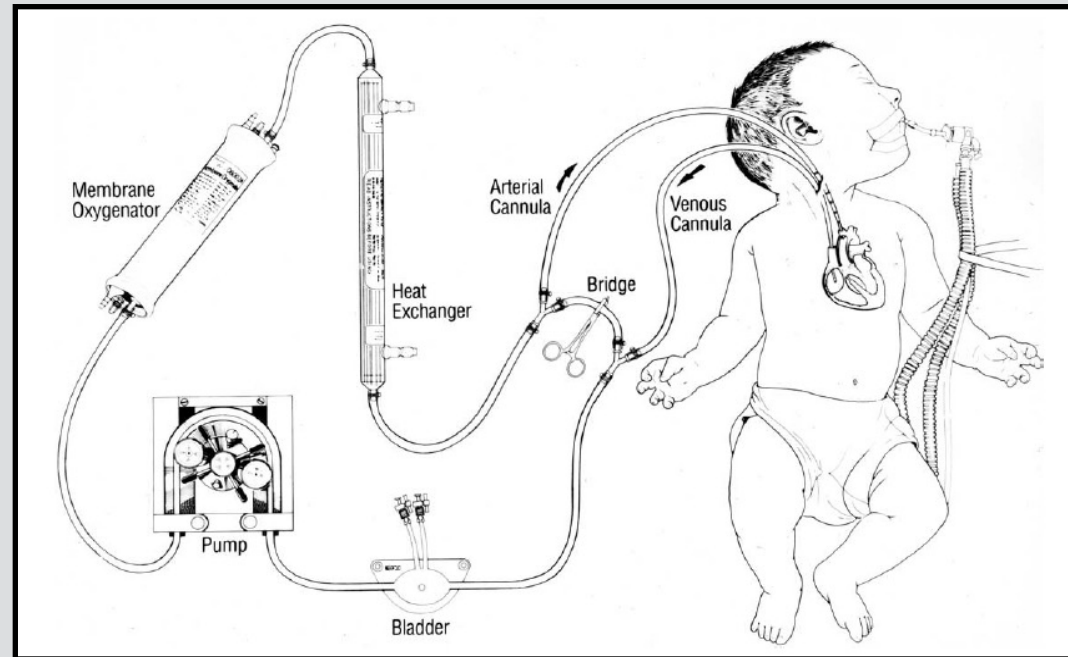


Assistance Circulatoire



Les assistances circulatoires

V-A ECMO support cardio-respiratoire



V-V ECMO support respiratoire



ECMO VV

- Jugulaire unique
- Jugulo-fémorale

ECMO VA

- Jugulo-carotidienne
- Fémoro-fémorale
- Centrale OD/Ao



Indications de l'ECMO

Défaillance post-cardiotomie

- Dysfonction myocardiaque pré-opératoire (ALCAPA, hypoVG, TGV vieillie)
- Dysfonction myocardiaque post-opératoire (clampage Ao prolongé, cardioprotection, dysfct VD, lésion résiduelle)
- Post-transplantation

Défaillance circulatoire « médicale »

- Myocardite, décompensation CMD
- Intoxication médicamenteuse
- Arythmies réfractaires
- ACR
- Hypoxémie et CHD



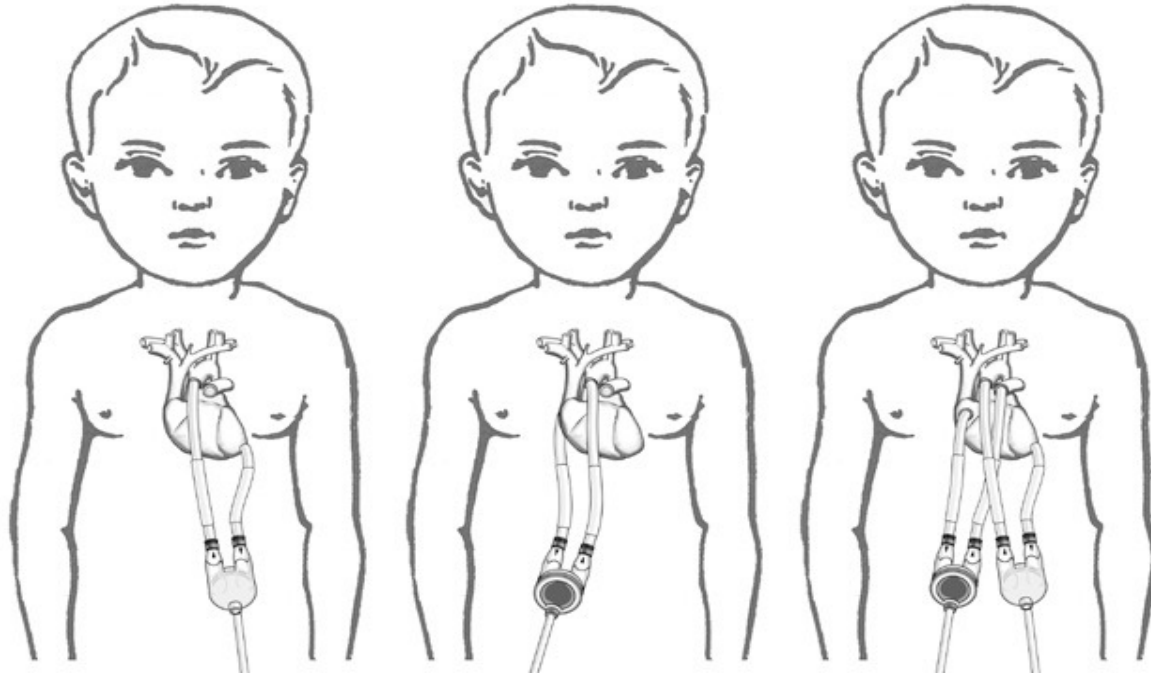
Should Extracorporeal Membrane Oxygenation Be Offered? An International Survey

Kevin W. Kuo, MD¹, Ryan P. Barbaro, MD¹, Samir K. Gadepalli, MD², Matthew M. Davis, MD³, Robert H. Bartlett, MD², and
Folafoluwa O. Odetola, MD¹

Contre- indications de l'ECMO

- Réanimation cardio-pulmonaire prolongée (+de 5 à 30 min dans les recommandations de l'ECLS0)
- Cardiopathie irréversible inopérable
- Atteinte neurologique sévère (acquise ou congénitale)
- Saignement actif important
- Terrain (sd polymalformatif, trisomie 18...)
- Nouveau né - de 2kg / - de 35 SA
- Canulation impossible sur un patient en arrêt

Assistance de longue durée - Berlin Heart



VAD
mono G
VG/Ao

VAD
mono D
OD/AP

biVAD

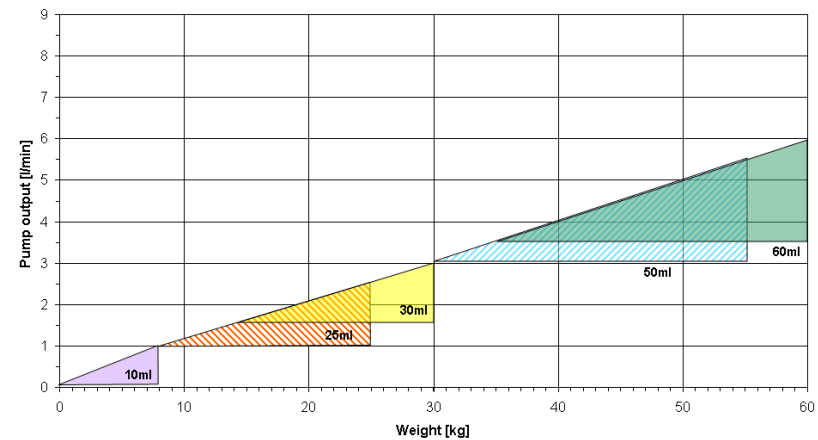


Berlin Heart

Ventricules de 10 à 80ml : patients de 3kg jusqu'aux adultes



Selection of pump size





Transplantation



Indications

En 2014, 586 Tx sur 112 centres (soit 5/centre/an)

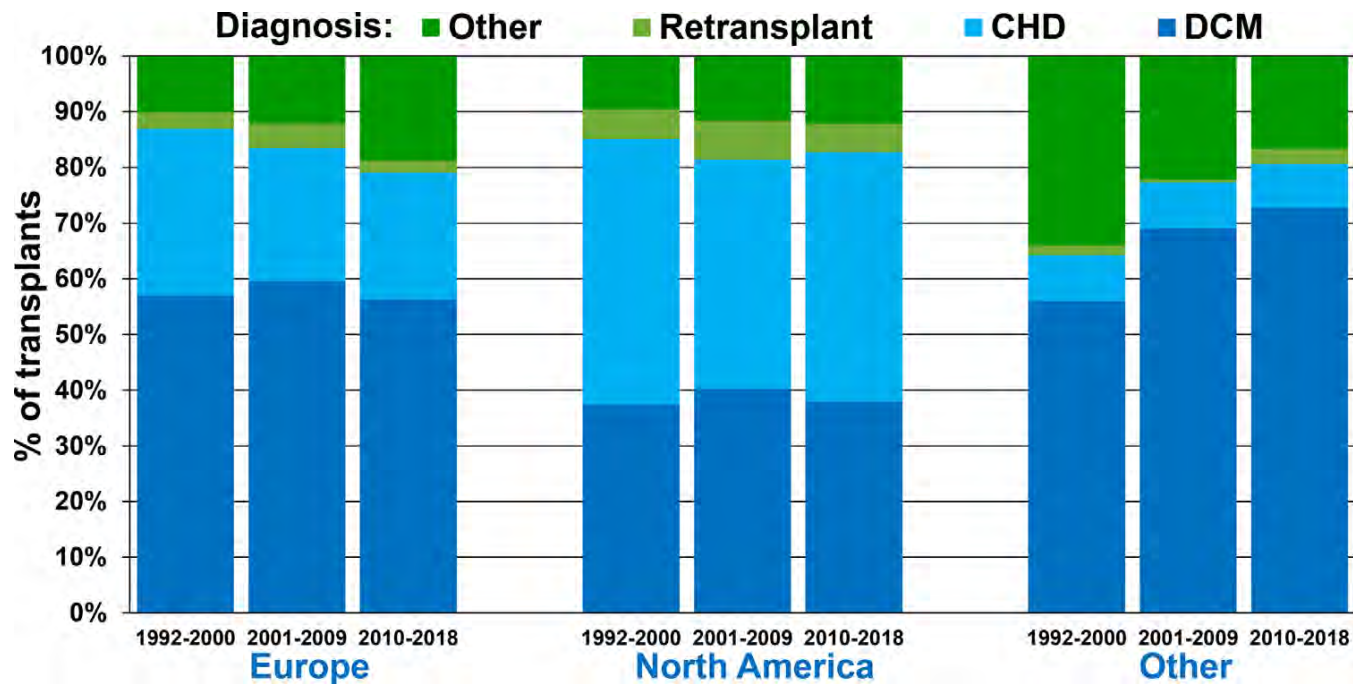


Figure 1 Recipient diagnosis by location and era (January 1992-June 2018).

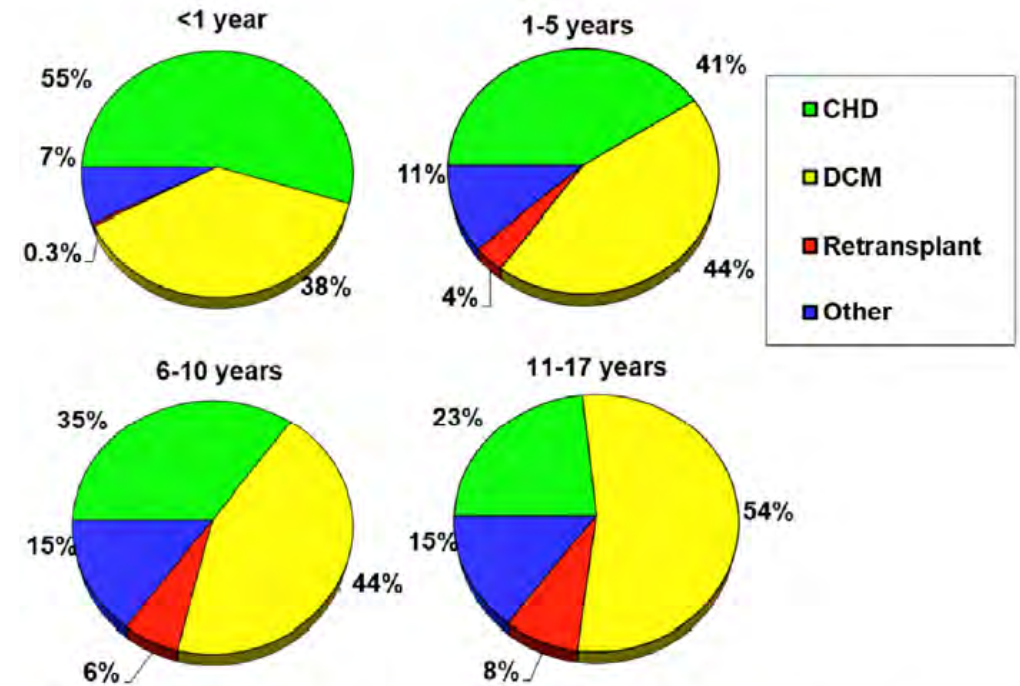


Figure 20 Recipient diagnosis by age group.

Statut des receveurs

Table 1 Distribution of Recipient Characteristics by Transplant Era (Transplants: January 1992 – June 2018)

	Jan 1992-Dec 2000(<i>n</i> = 3,666)	Jan 2001-Dec 2009(<i>n</i> = 4,476)	Jan 2010-Jun 2018(<i>n</i> = 5,307)	<i>p</i> -value
Age (years)	6 (0 - 17)	7 (0 - 17)	7 (0 - 17)	0.0005
Age <1 year	25.5%	22.7%	21.9%	0.0003
Male	58.2%	54.6%	54.5%	0.0009
BMI (kg/m ²)	19.9 (16.1 - 30.0)	20.9 (16.3 - 32.4)	21.1 (16.4 - 31.7)	<0.0001
PRA ≥20%	6.7%	15.1%	23.8%	<0.0001
PRA ≥80%	1.3%	5.1%	5.9%	<0.0001
History of malignancy	3.2% ^a	2.4%	2.2%	0.0653
Pre-transplant dialysis	1.9% ^a	4.1%	3.3%	0.0002
Bilirubin (mg/dl)	0.8 (0.2 - 3.2) ^a	0.7 (0.2 - 3.4)	0.6 (0.2 - 2.9)	<0.0001
Creatinine (mg/dl)	0.6 (0.3 - 1.3) ^a	0.5 (0.2 - 1.4)	0.4 (0.2 - 1.1)	<0.0001
GFR (ml/min/1.73 m ²) ^c	80.7 (39.6 - 127.2) ^a	82.6 (39.4 - 138.8)	94.0 (50.8 - 164.2)	<0.0001
PVR (woods unit)	3.0 (0.8 - 11.8) ^a	3.0 (0.8 - 12.2)	2.6 (0.6 - 11.8)	0.0046
Inotrope use	42.2% ^a	49.6%	50.5%	<0.0001
PGE use, age <1 year ^d	32.2% ^b	11.6%	8.7%	<0.0001
ECMO use	4.4% ^b	8.6%	4.4%	<0.0001
MCS use:				
- None	-	86.3%	74.3%	<0.0001
- VAD	-	9.8%	20.0%	
- TAH	-	0.2%	0.3%	
- BIVAD	-	3.7%	5.4%	
Ventilator use	17.4%	21.2%	16.6%	<0.0001
Hospitalized	64.7%	69.3%	72.2%	<0.0001

Le donneur idéal

CONDITIONS GENERALES

- Age < 40-50 ans
- Pas de tabac ou <10-20 PA
- Pas d'ATCD de néoplasie, traumatisme thoracique, chirurgie cardio-thoracique
- Distance (ischémie)

CONDITIONS SPECIFIQUES

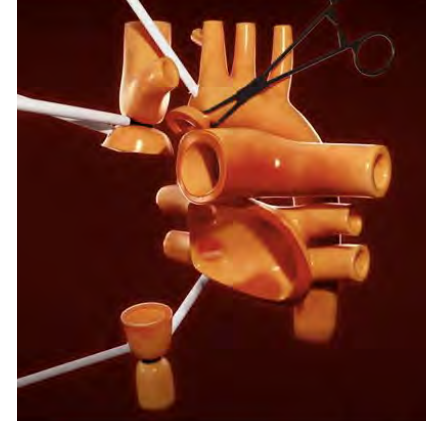
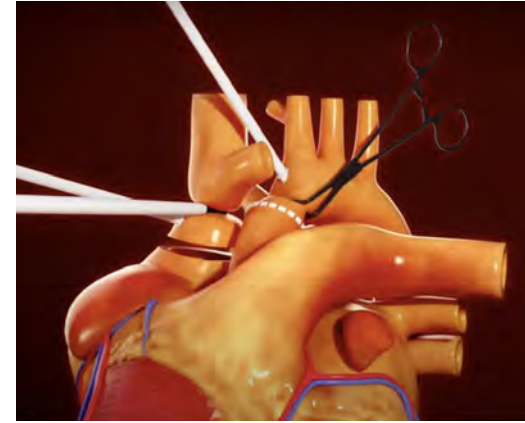
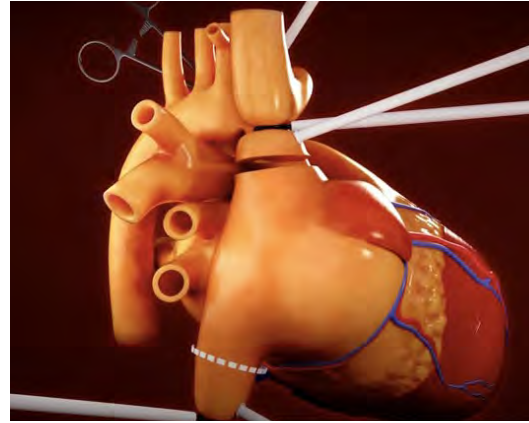
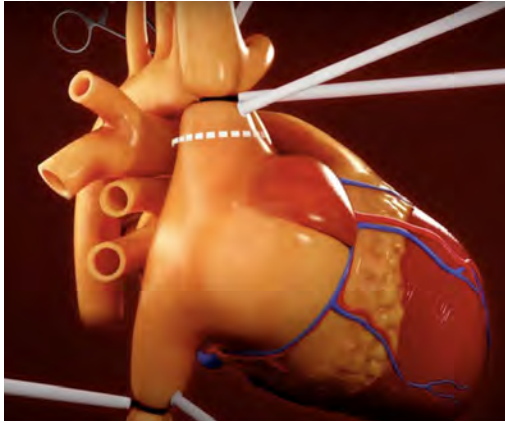
- Pas ou peu d'amines
- Pas ou peu de troponine
- ETT normale
- Coronaires saines



Choix du greffon : selon le patient

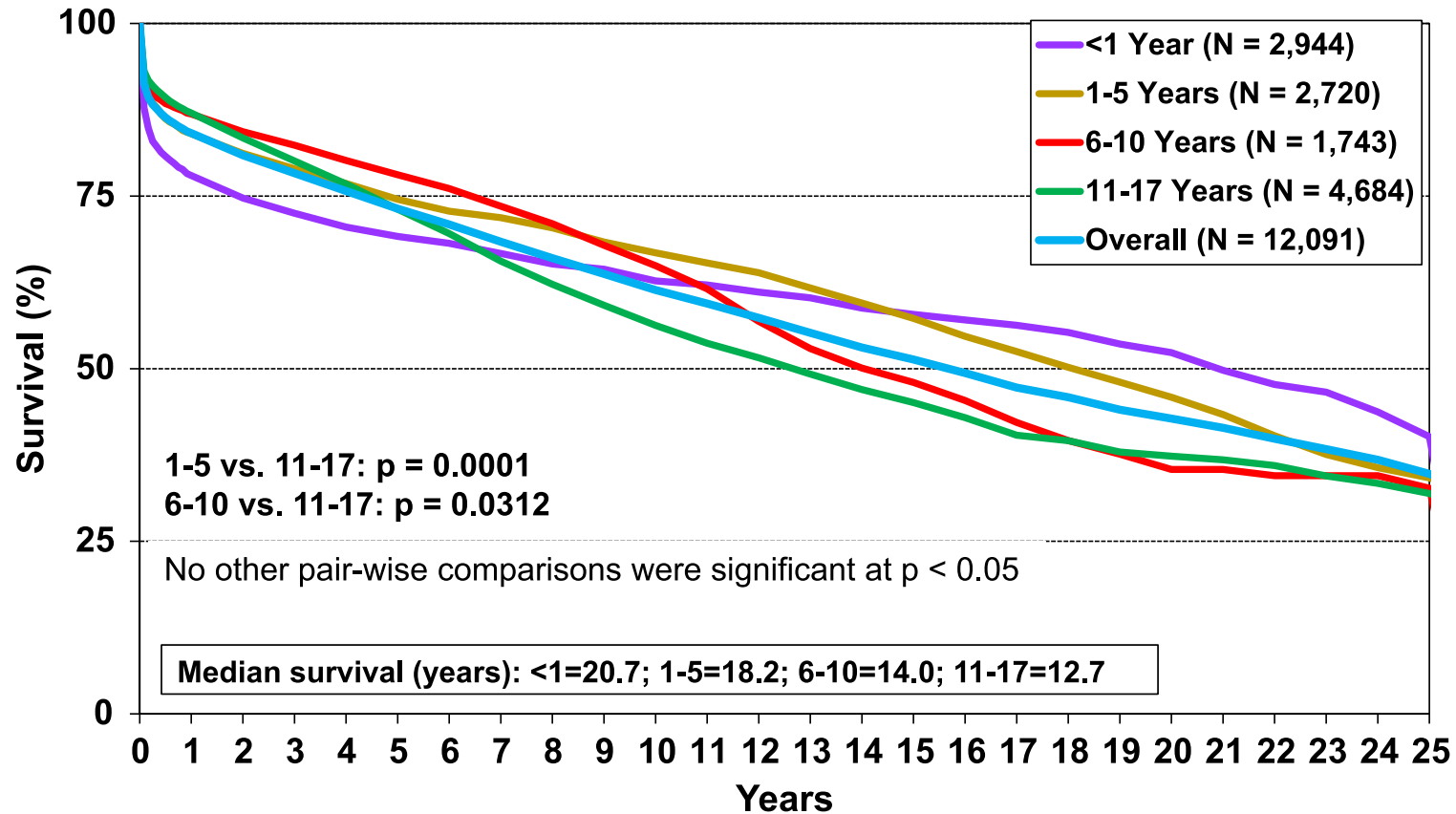
- CRITERES GENERAUX
 - Groupe sanguin
 - CMV et EBV
 - Antigènes interdits
- CRITERES SPECIFIQUES
 - Morphologie – Poids
 - Indication - Poids

Explantation cardiaque receveur



<https://www.youtube.com/watch?v=VGZRMHA4ics>

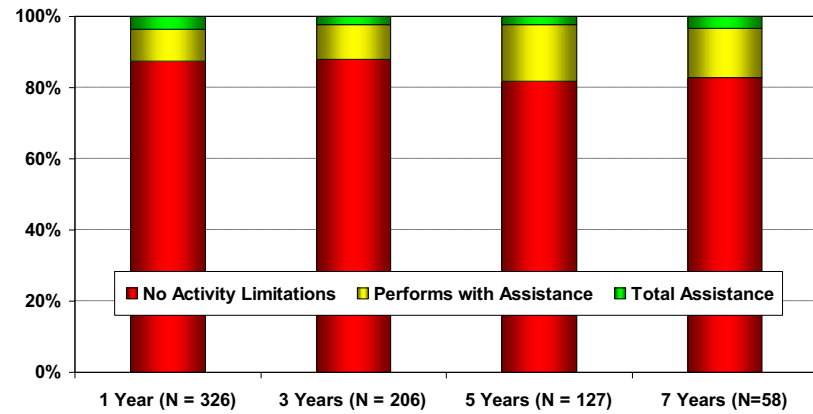
Survie après Transplantation cardiaque



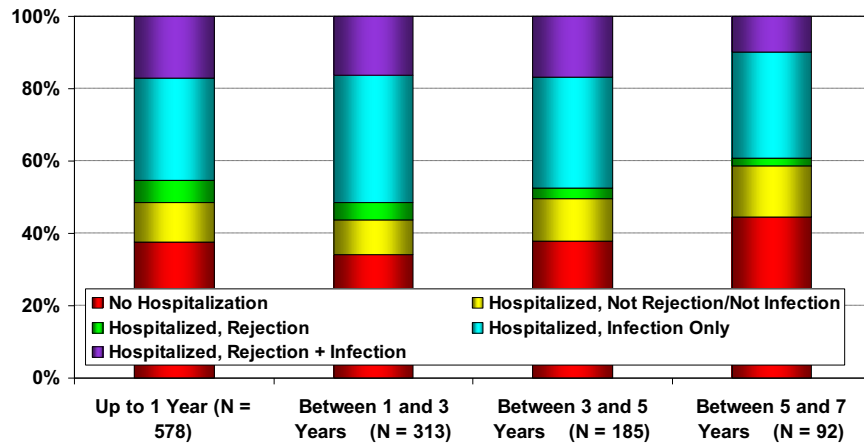
Médiane de survie de 15 ans

Comment vont-ils ?

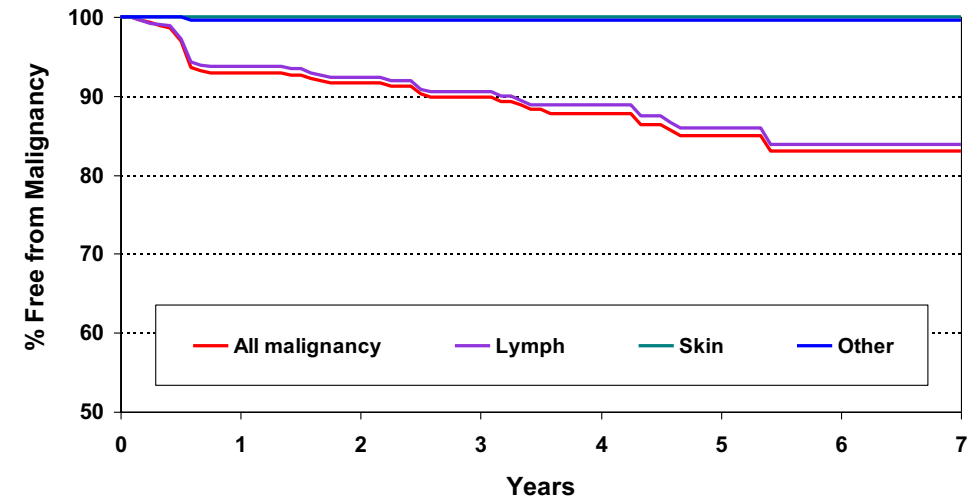
Statut fonctionnel



Réhospitalisation



Morbi-mortalité



Alternative ?

Cerclage prophylactique et cardiomyopathie dilatée

Early prophylactic pulmonary artery banding in isolated congenitally corrected transposition of the great arteries[☆]

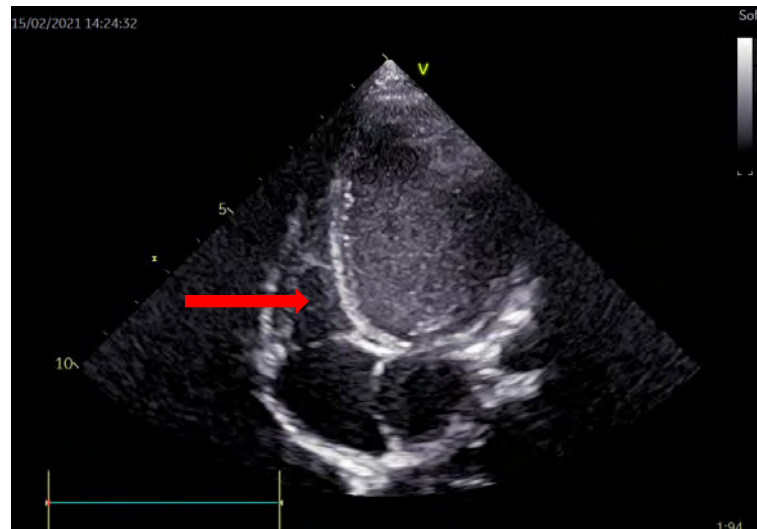
European Journal of Cardio-thoracic Surgery 38 (2010) 728–734

Pulmonary artery banding in infants and young children with left ventricular dilated cardiomyopathy: A novel therapeutic strategy before heart transplantation

The Journal of Heart and Lung Transplantation, Vol 32, No 5, May 2013

Cerclage prophylactique

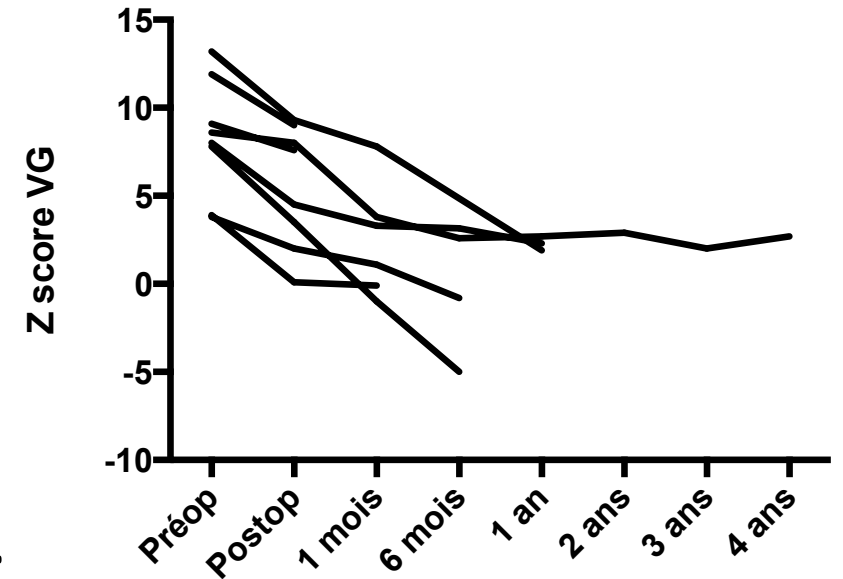
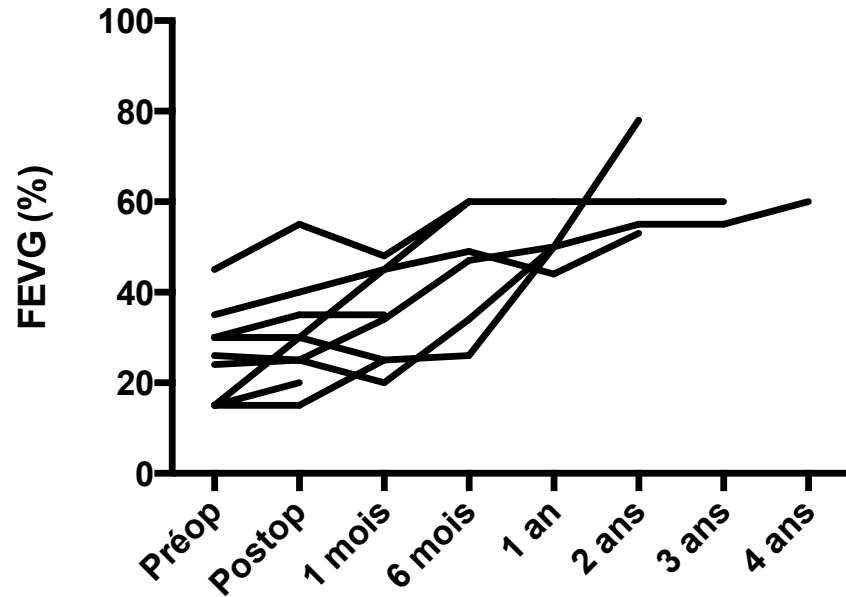
- Monitoring ETO en continu
- Incision limitée du péricarde
- Cerclage sous Adrénaline et Milrinone
- Objectif PVD 60-70%



Cerclage prophylactique

FEVG :

- 26% avant cerclage
- 50% (+ 22.6%) au dernier suivi



DTDVG :

- 44.5mm avant cerclage
- 38 mm (- 6.4 mm) au dernier suivi

Z score :

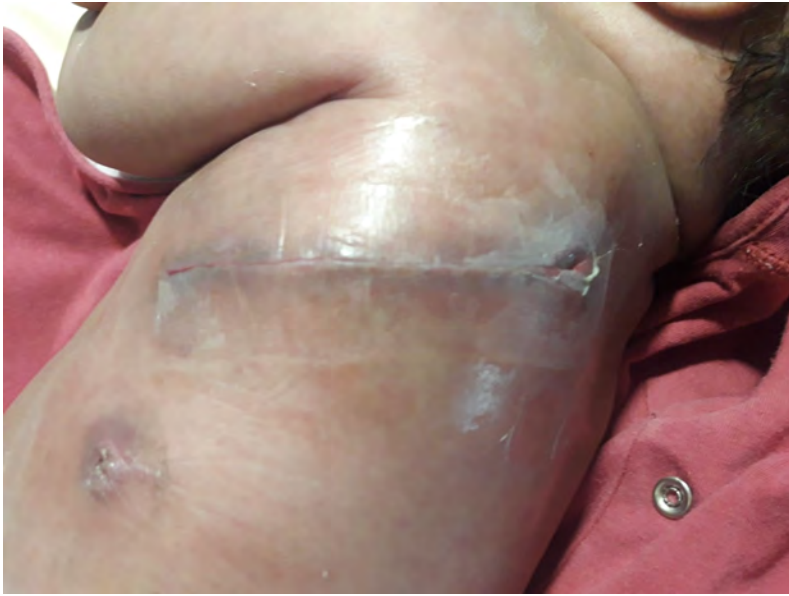
- + 8.5 avant cerclage
- + 1.8 (- 6) au dernier suivi



Cicatrices



Cicatrice de Thoracotomie



Cicatrice de Thoracotomie



Cicatrice de Sternotomie



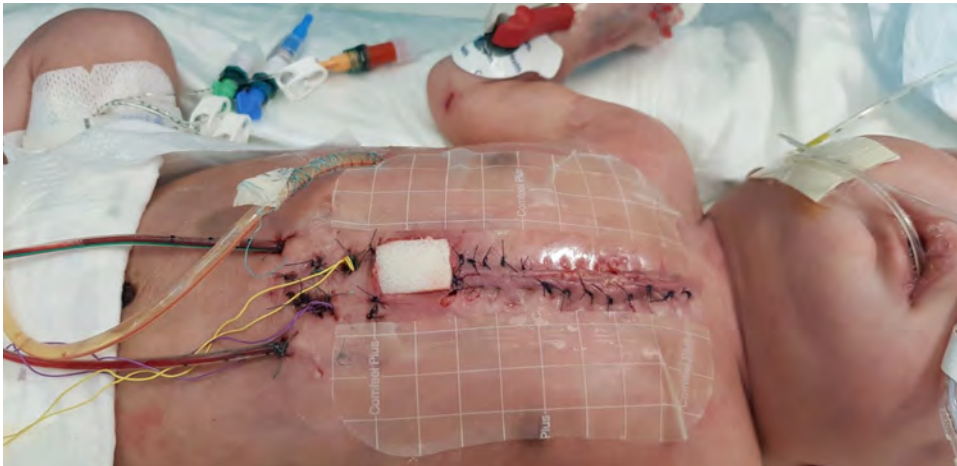
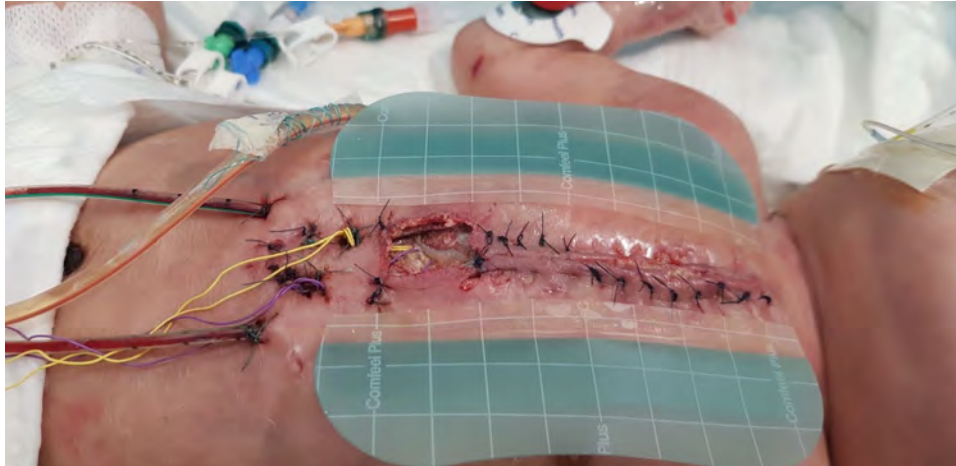
Cicatrice de Sternotomie



Cicatrice de Sternotomie



Cicatrice de Sternotomie



Merci de votre attention
margaux.pontailier@aphp.fr

