



Cardiopathies congénitales

Simple et complexes en même temps

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Hôpital Universitaire Necker Enfants malades – APHP, Université Paris Descartes, Sorbonne Paris Cité
IcarP Cardiology, Institut Hospitalo-Universitaire IMAGINE

Centre de Référence Maladies Rares
Malformations Cardiaques Congénitales Complexes-M3C
Centre de Référence Maladies Rares
Maladies Cardiaques Héritaires- CARDIOGEN

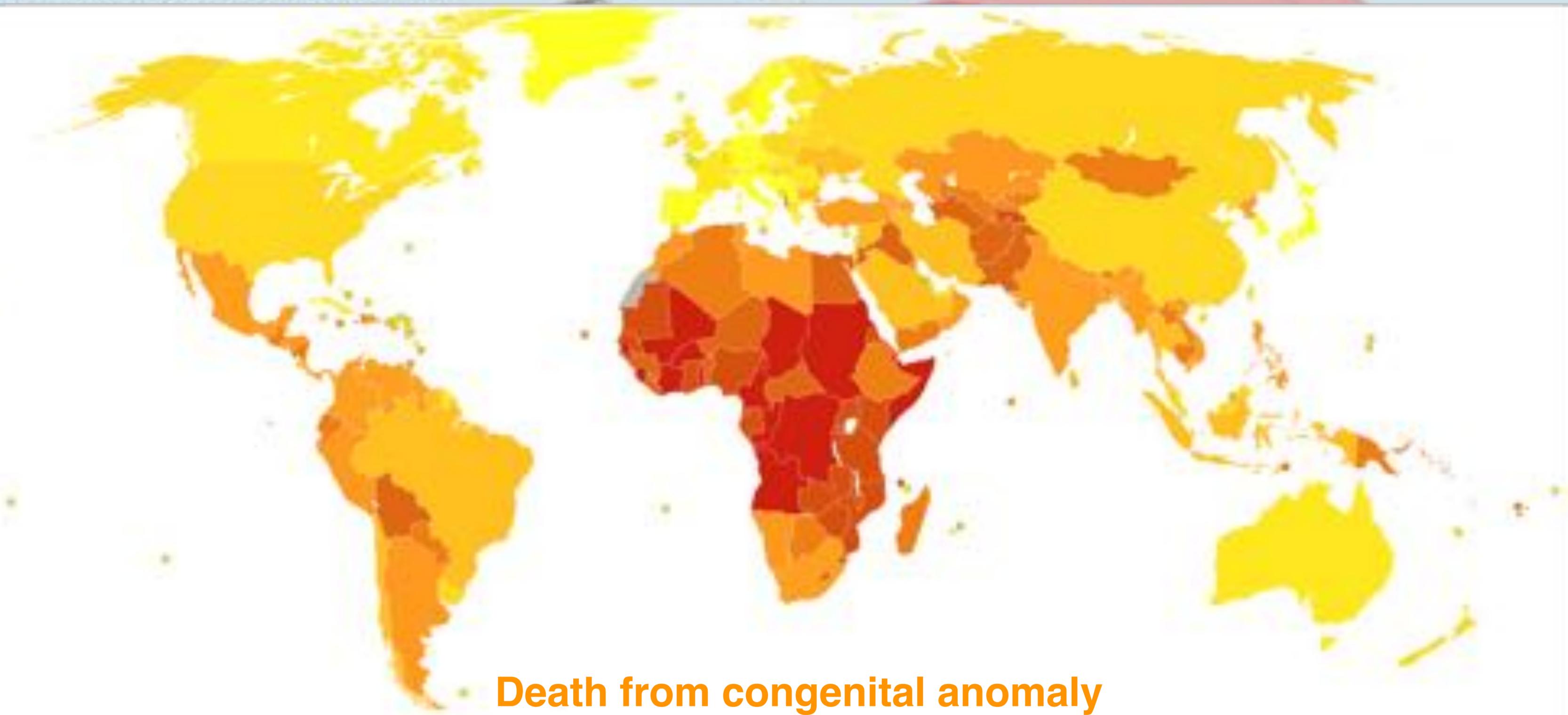




The landscape of congenital heart diseases



Multidimensional Poverty



Death from congenital anomaly

Le réseau M3C



Fédération des Cardiopathies Congénitales de l'Assistance Publique des Hôpitaux de Paris

Cardiologie Foetale



Cardiologie Congénitale et Pédiatrique



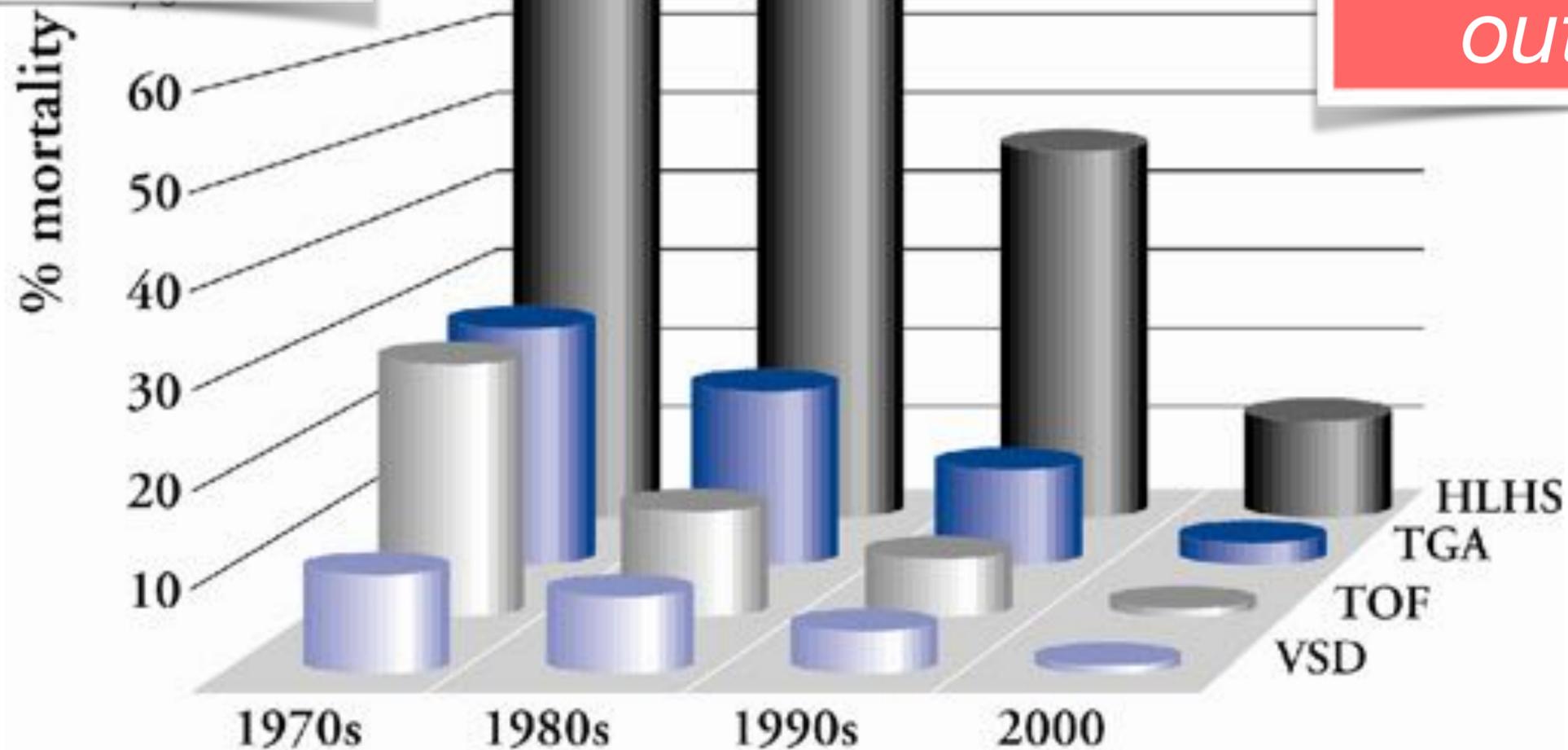
Cardiologie Congénitale Adulte



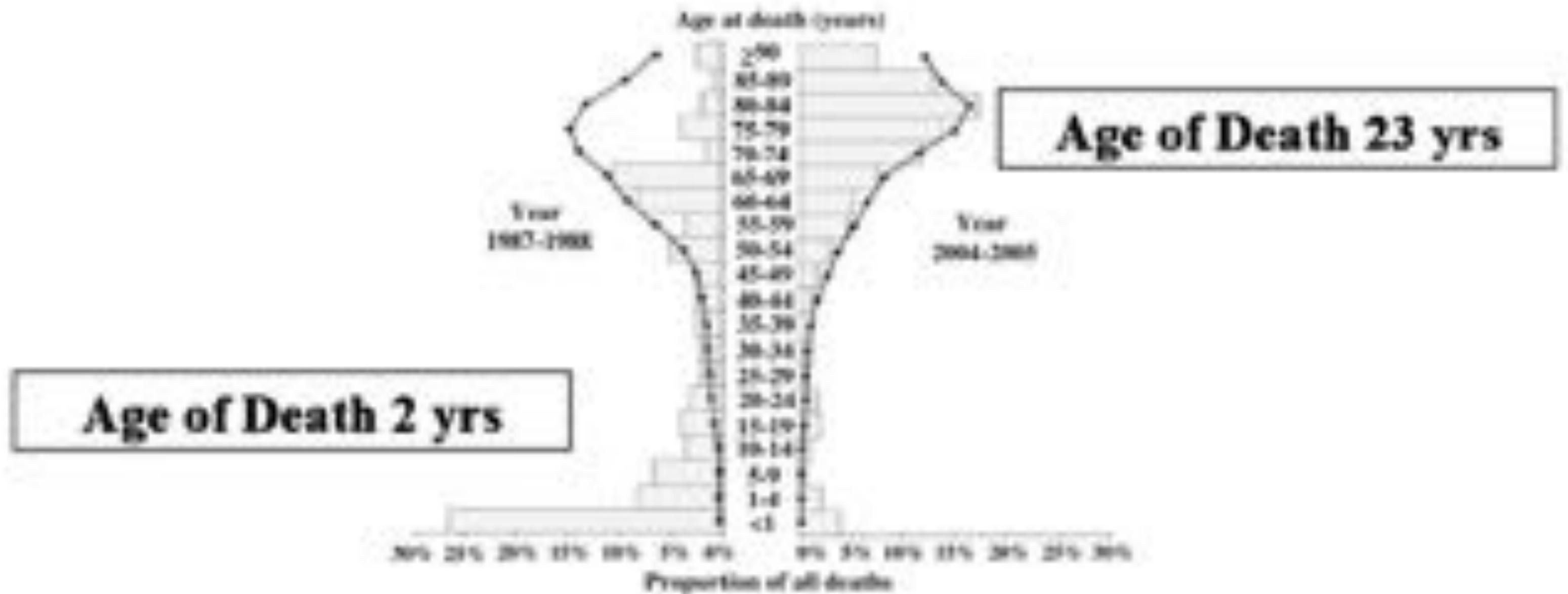
Improving Surgical Results: Mortality in CHDs

*Patient Survival
is Now
Expected*

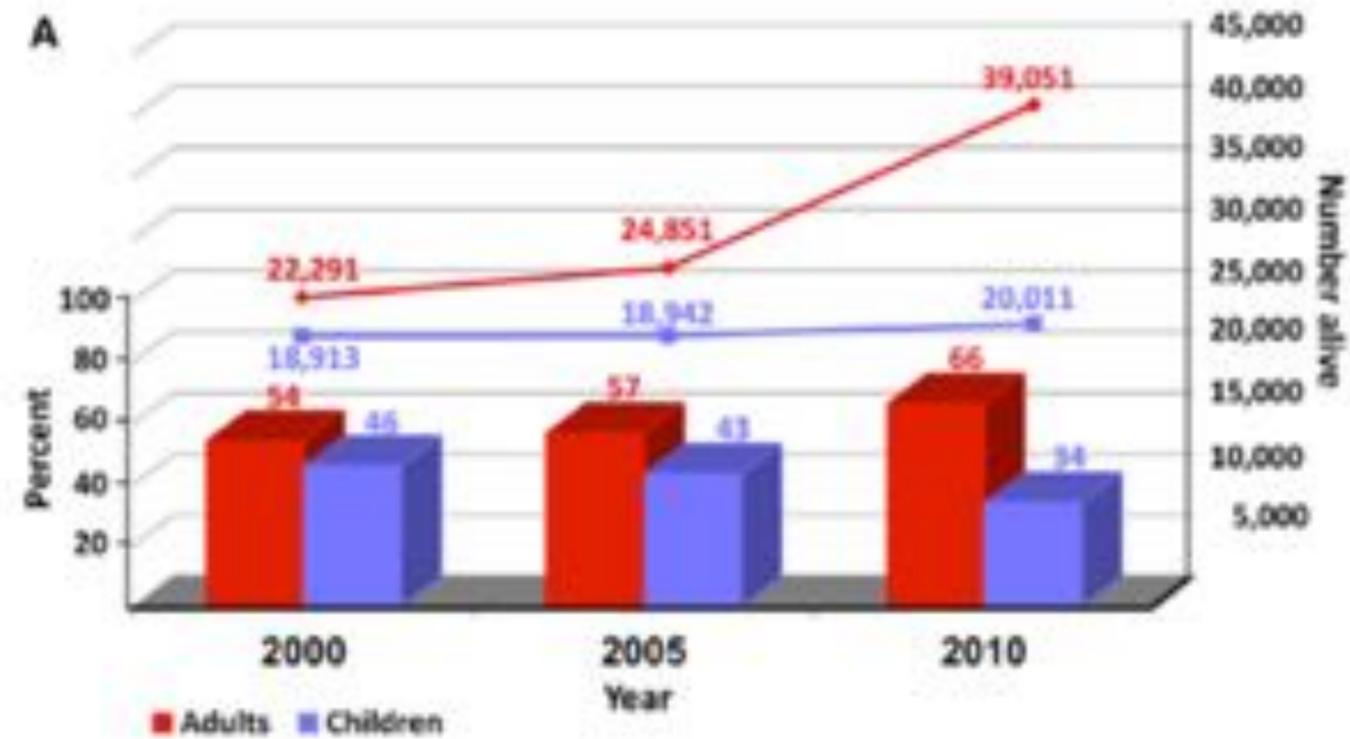
*In 2019, we
need to
redefine
outcomes*



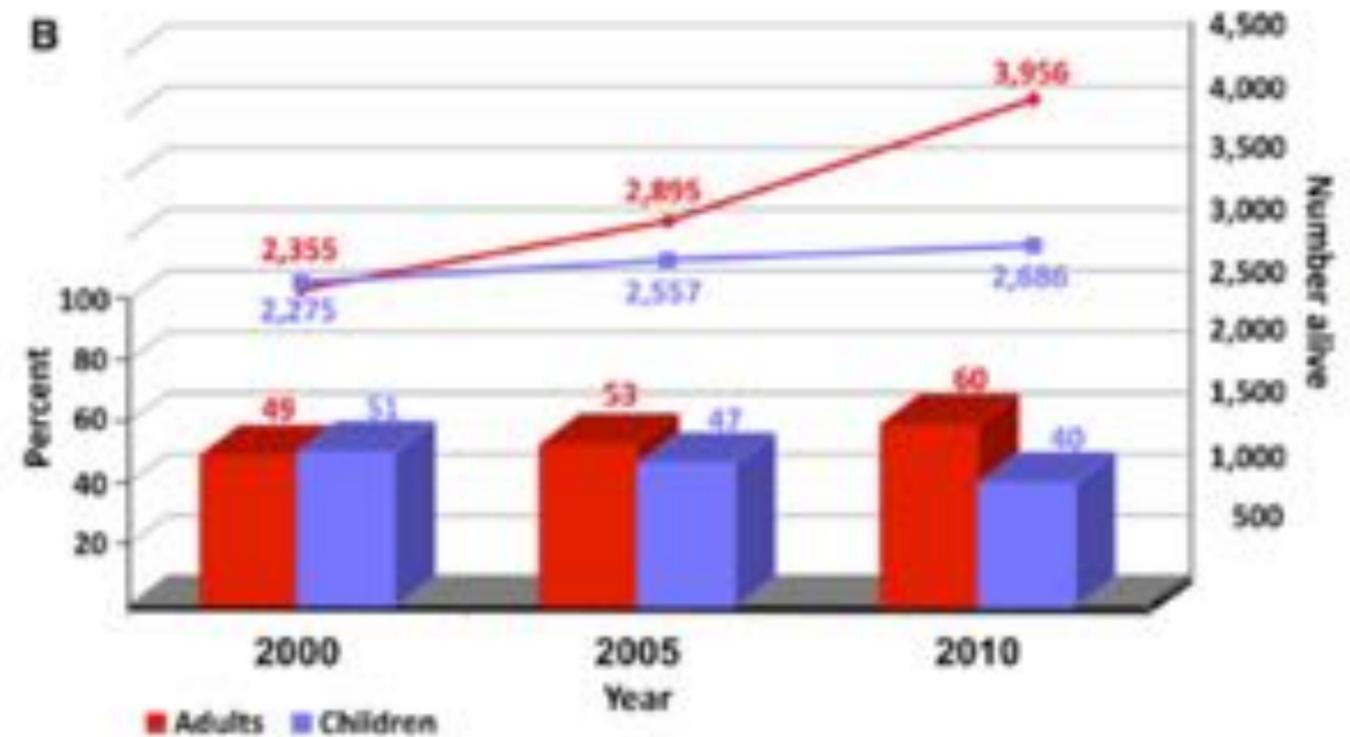
Evolution of mortality in CHD



Adults with CHD now outnumber their pediatric counterparts



Simple CHD

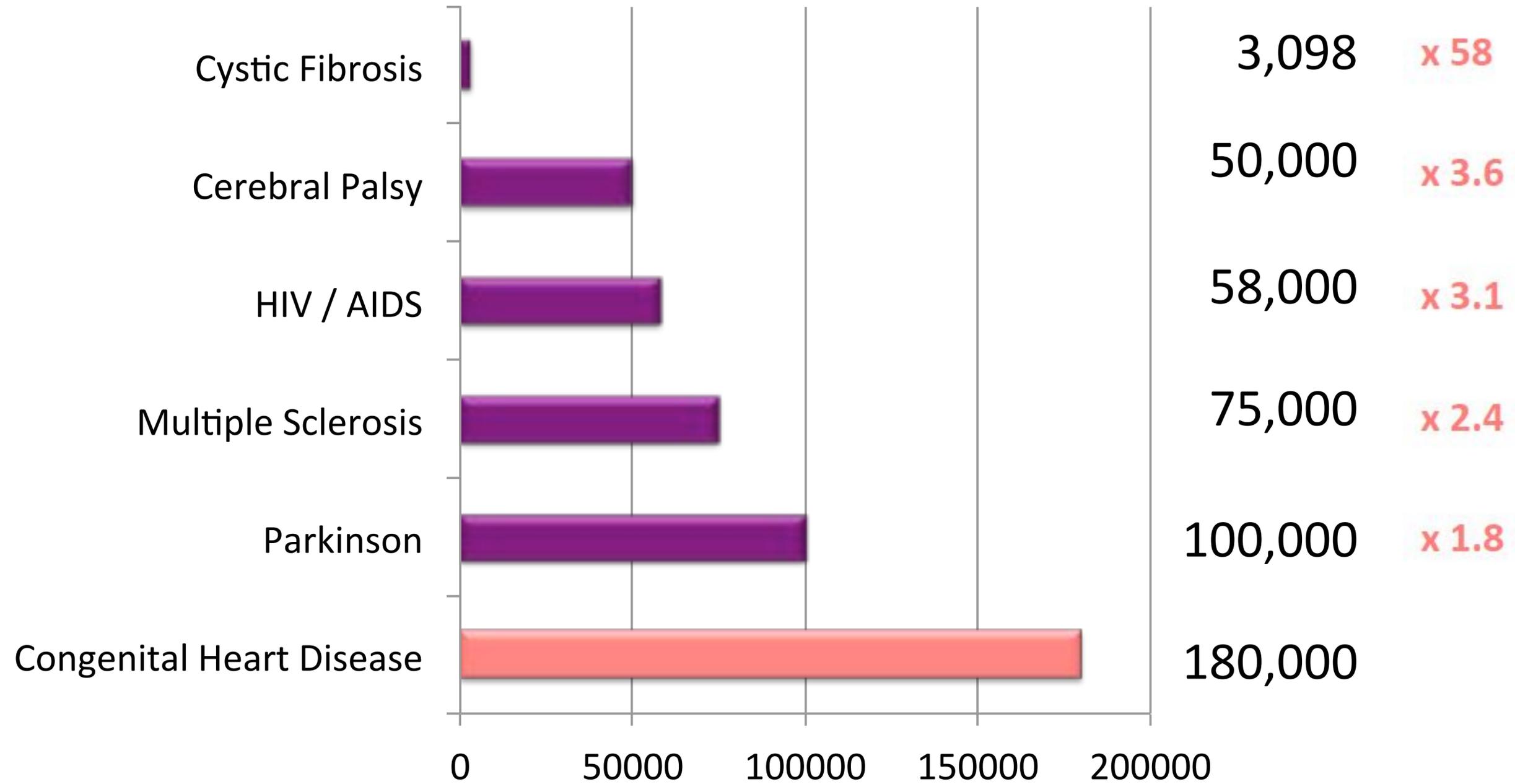


Complex CHD



Epidemiology of congenital heart diseases

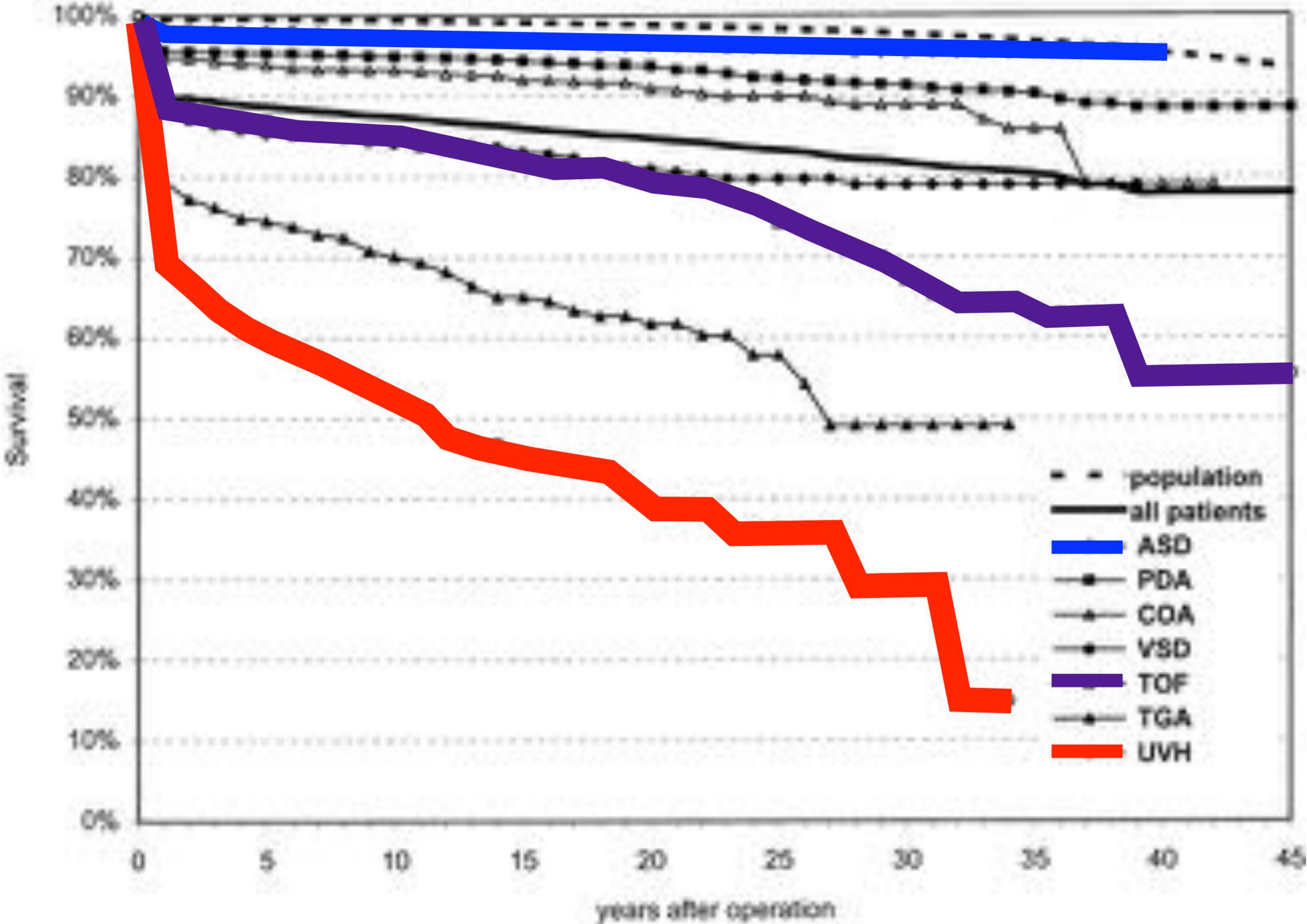
Comparison with other common diseases: Adult CHD is a public health problem



Sources: Cerebral Palsy Canada, Cystic Fibrosis Association, Public Health Agency of Canada; Canadian Congenital Heart Alliance



Mortality in Adult CHD according to the type of defect



Associations
extra cardiaques

Anatomie de la
malformation

Risque de
réintervention

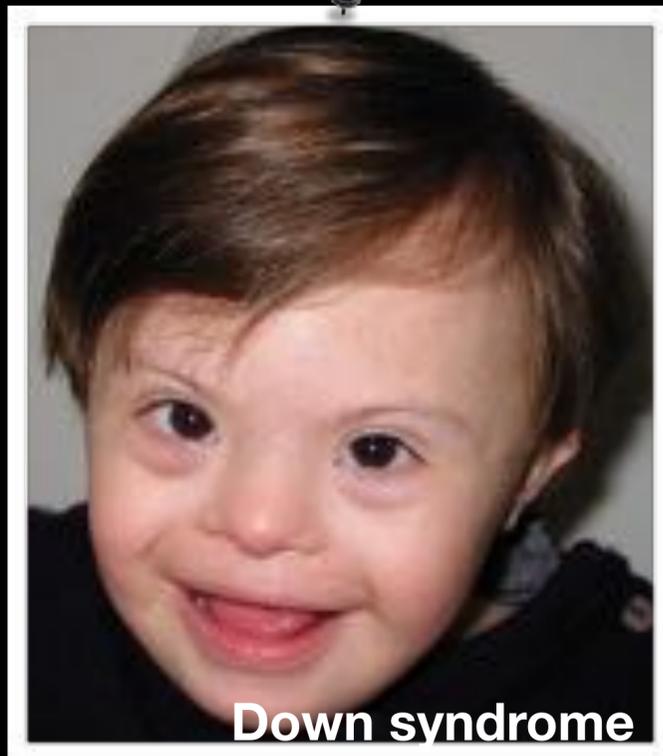
COMPLEX

SIMPLE

Timing
chirurgical

Réparée
ou non

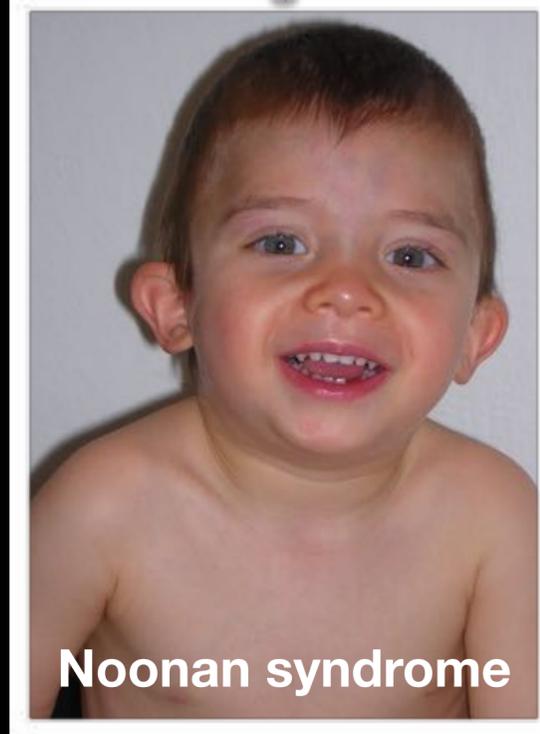
Associations extra cardiaques



Down syndrome



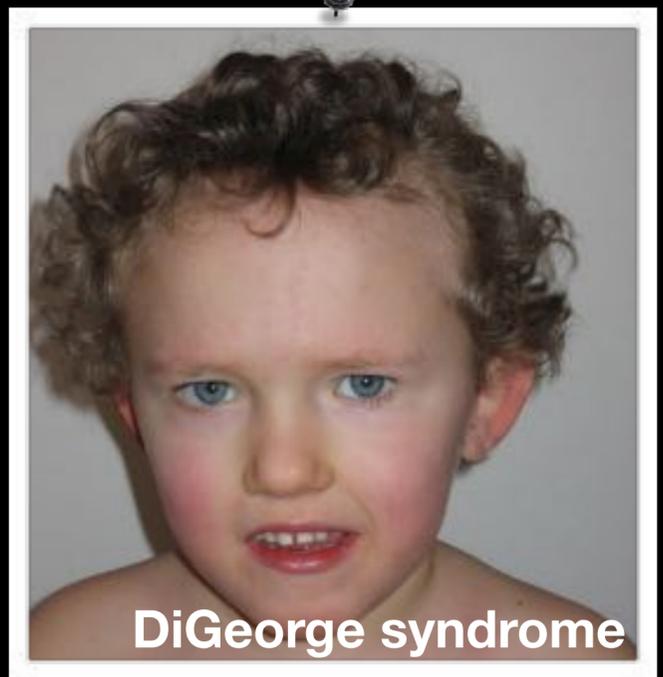
Turner syndrome



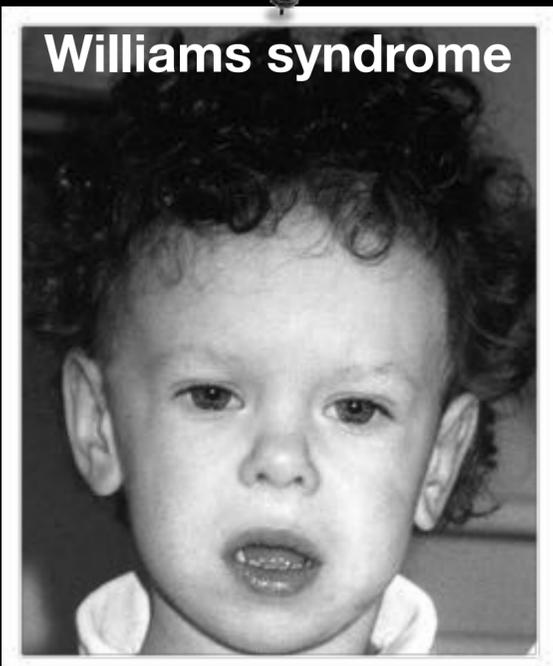
Noonan syndrome



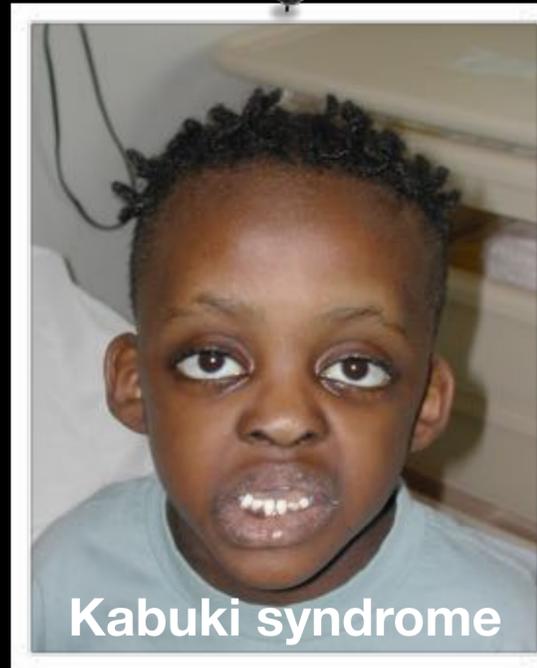
Marfan syndrome



DiGeorge syndrome



Williams syndrome

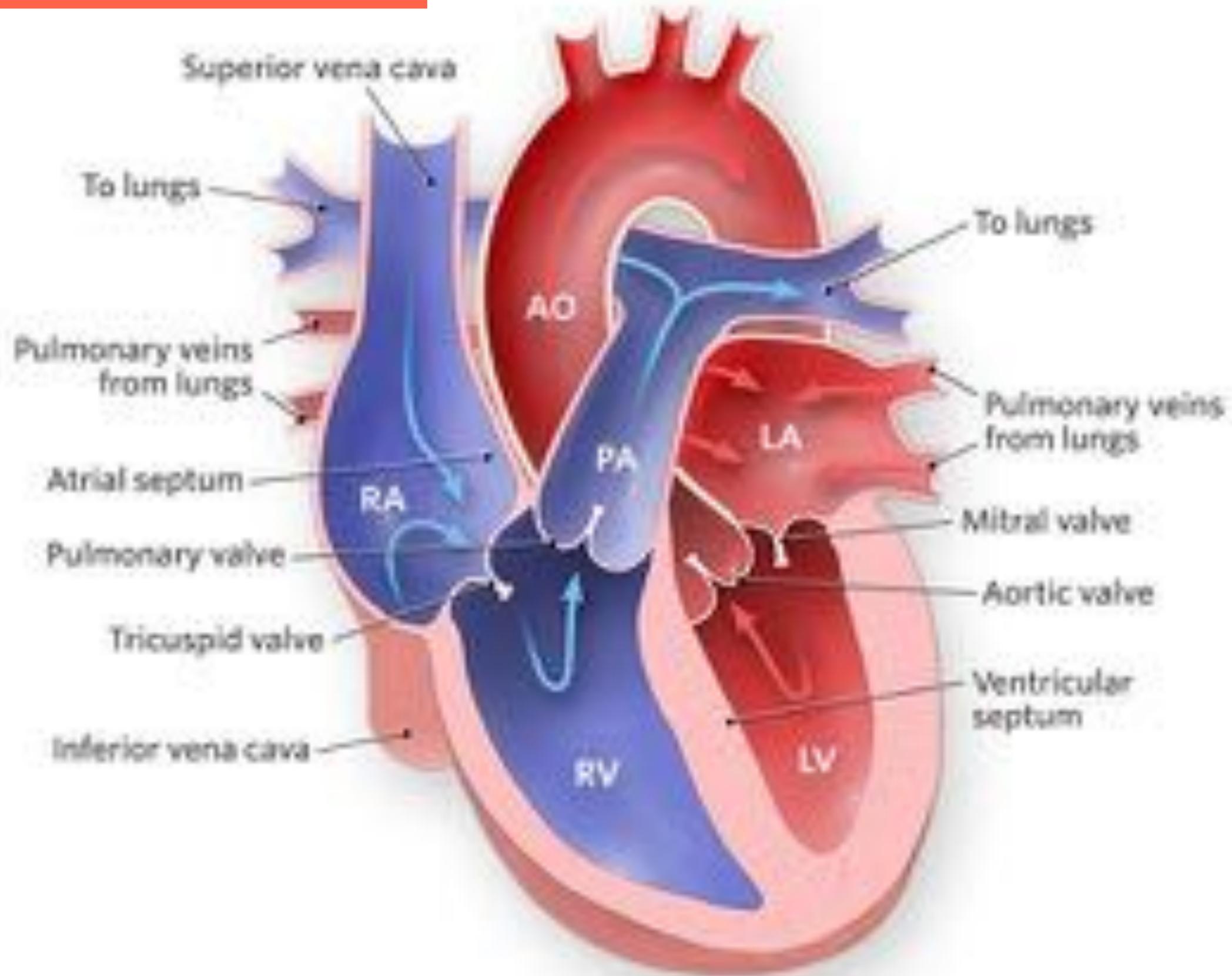


Kabuki syndrome

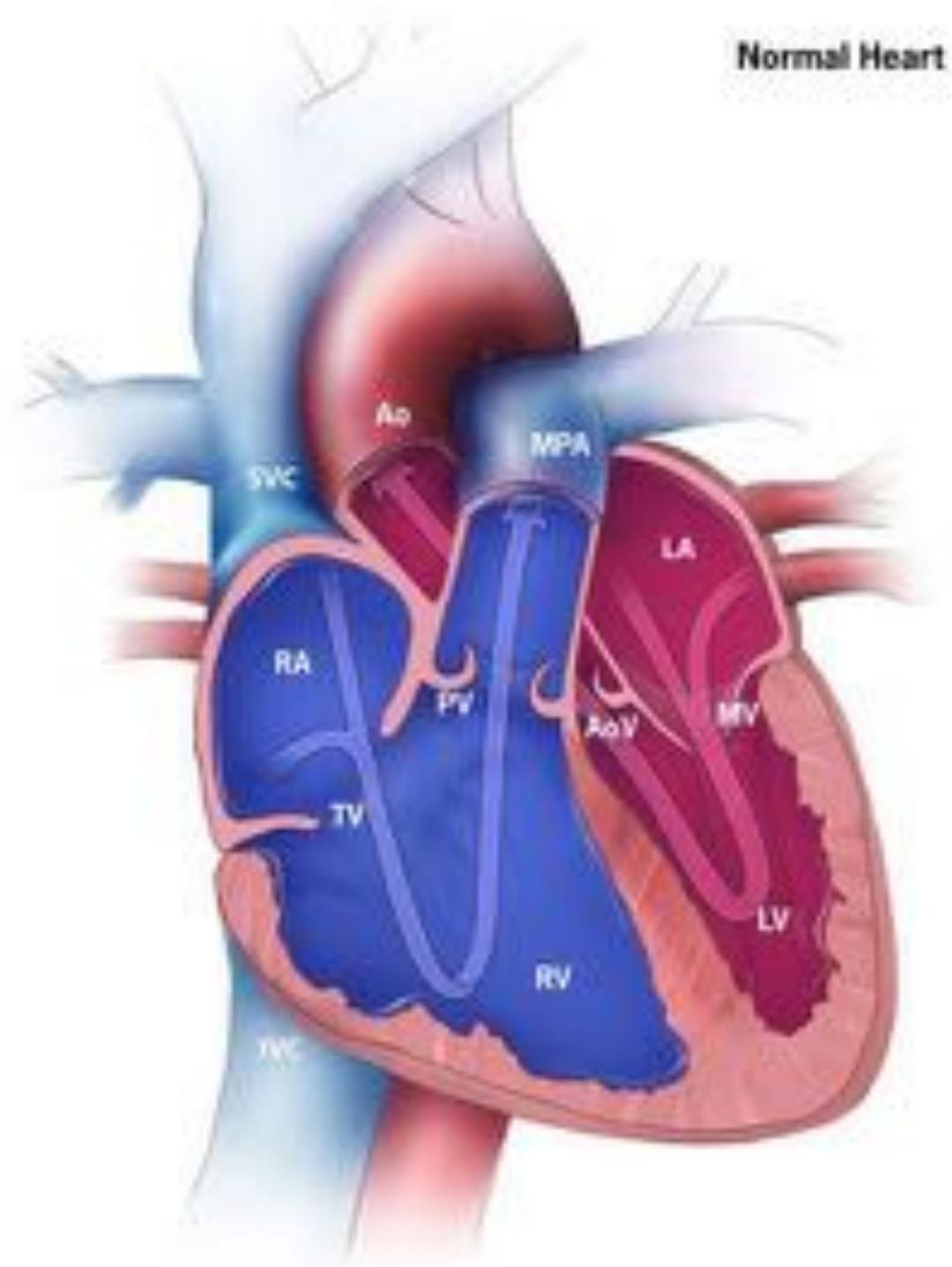


Wardenburg syndrome

Anatomie de la malformation



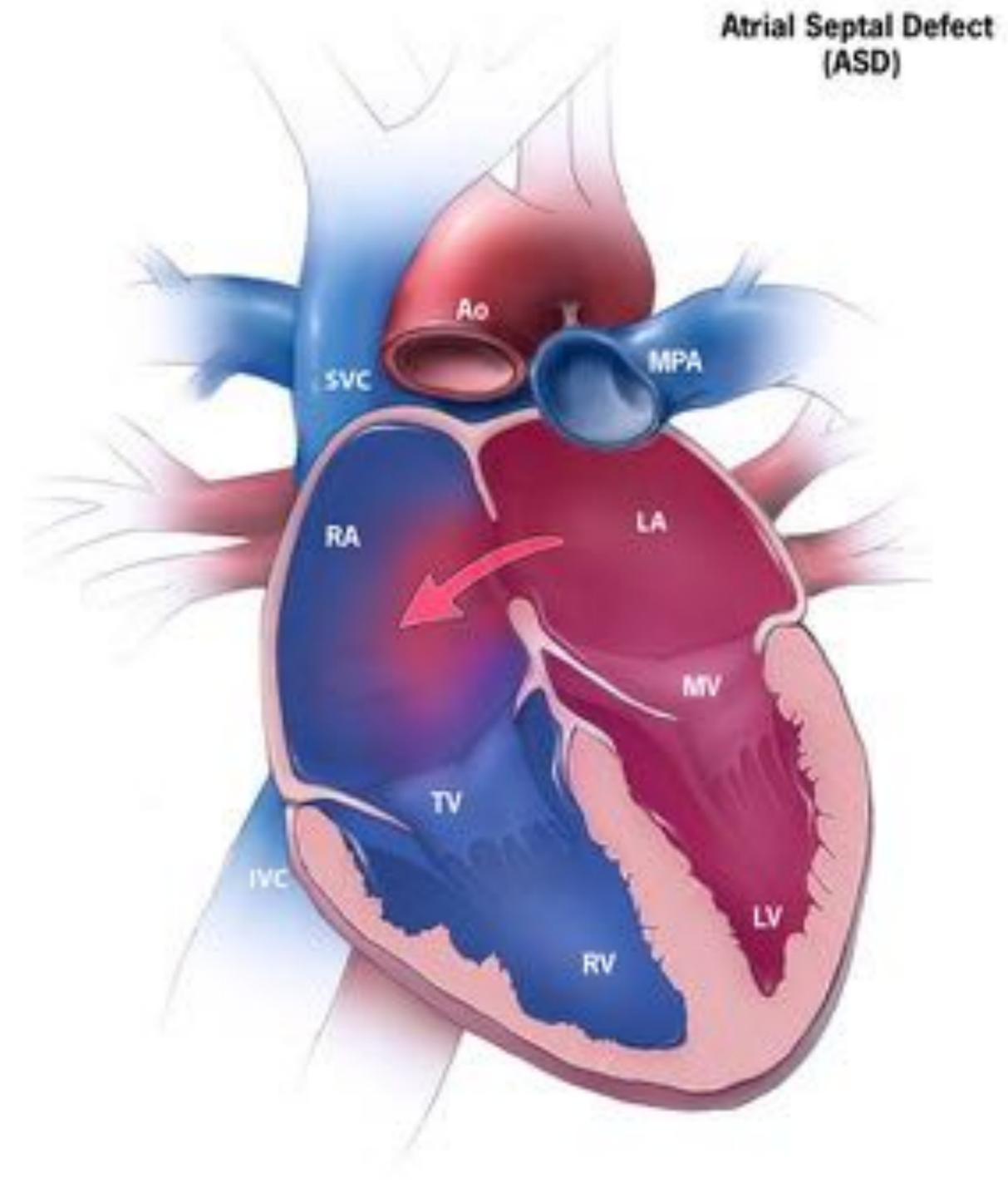
Cardiopathies simples



RA, Right Atrium
RV, Right Ventricle
LA, Left Atrium
LV, Left Ventricle

SVC, Superior Vena Cava
IVC, Inferior Vena Cava
MPA, Main Pulmonary Artery
Ao, Aorta

TV, Tricuspid Valve
MV, Mitral Valve
PV, Pulmonary Valve
AoV, Aortic Valve

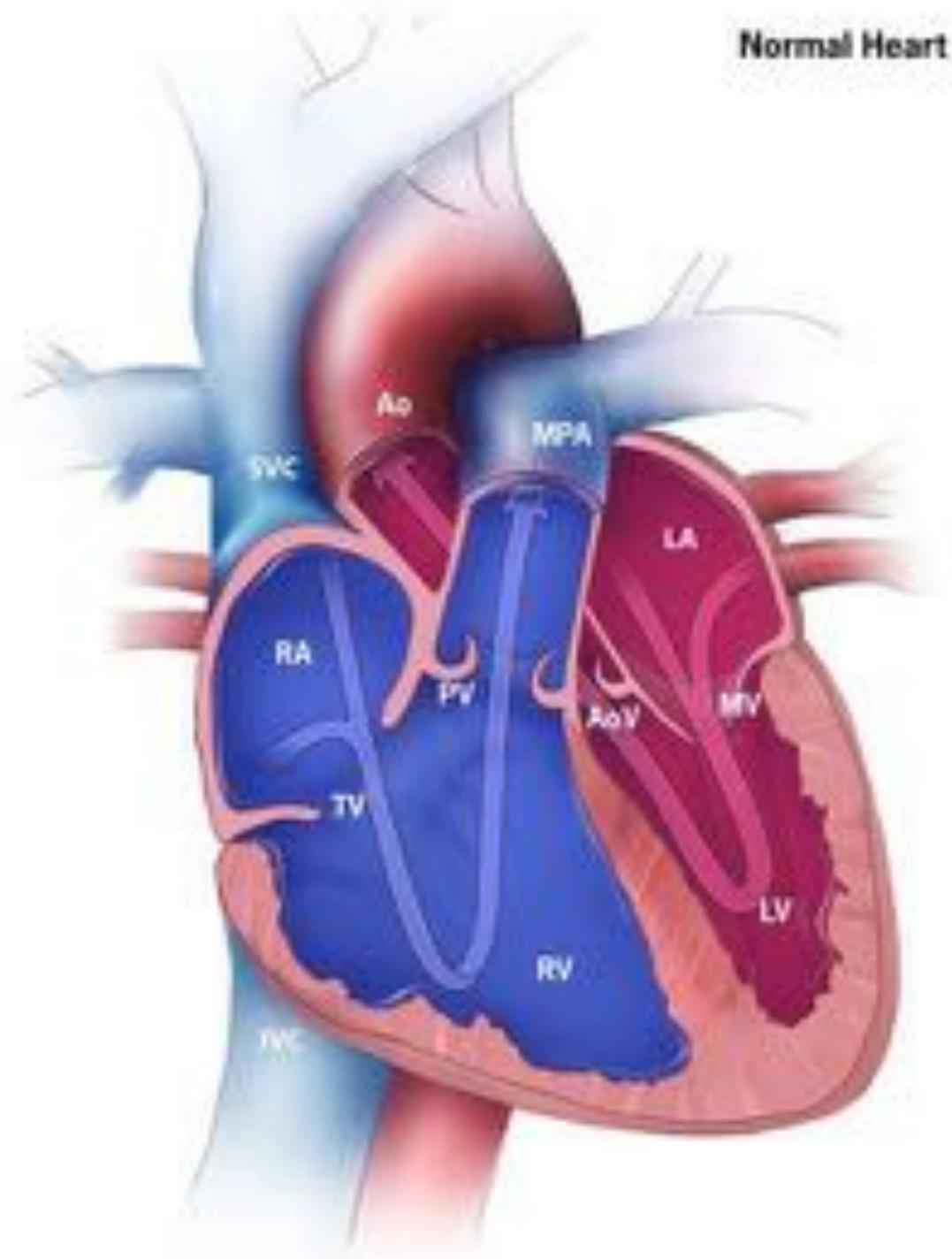


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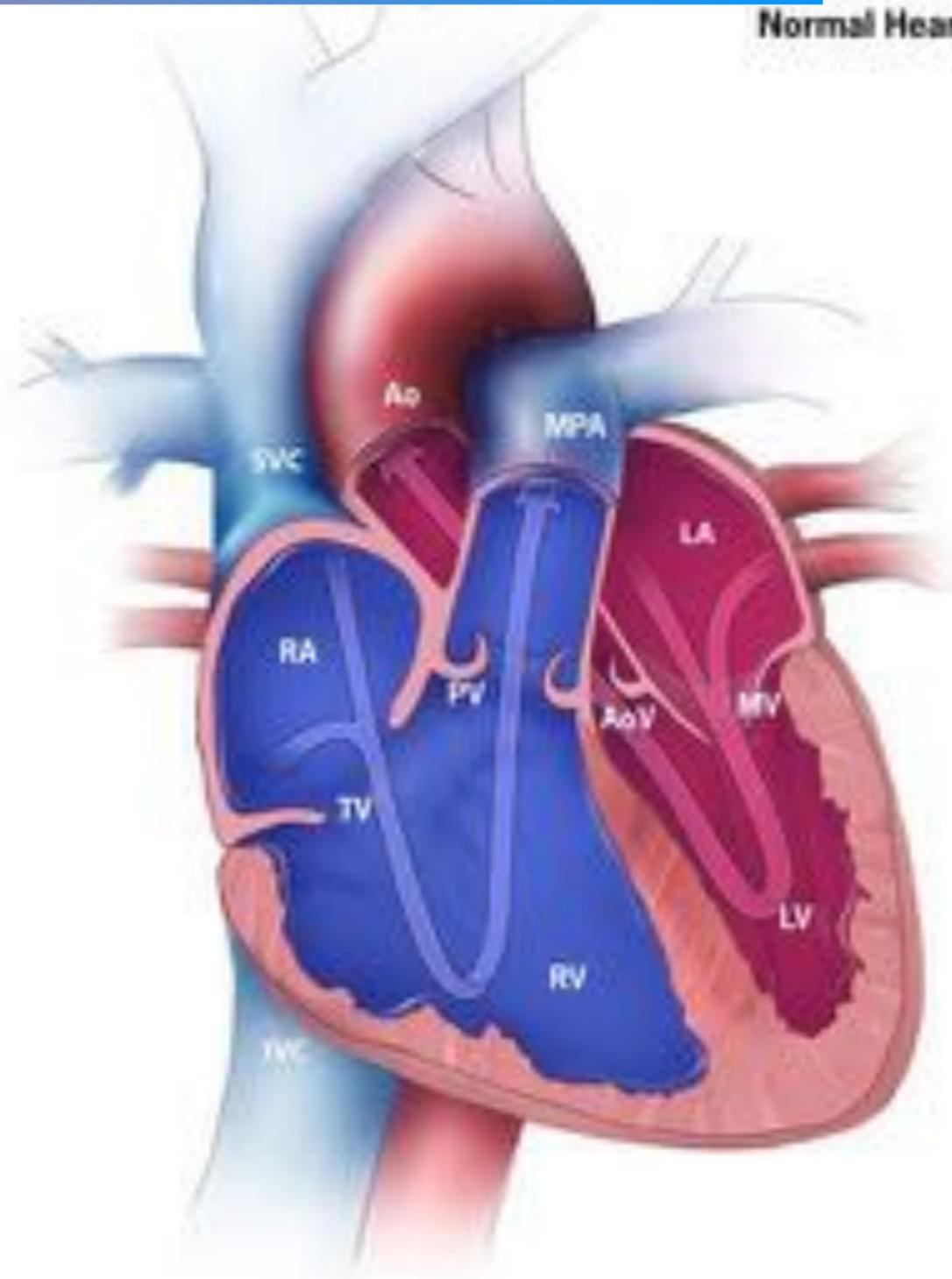
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1. Interventricular septum
2. Interventricular septum
3. VSD
4. Interventricular septum

Cardiopathies simples

Normal Heart



Interatrial Septal Defect (IASD)



RA, Right Atrium
RV, Right Ventricle
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LV, Left Ventricle

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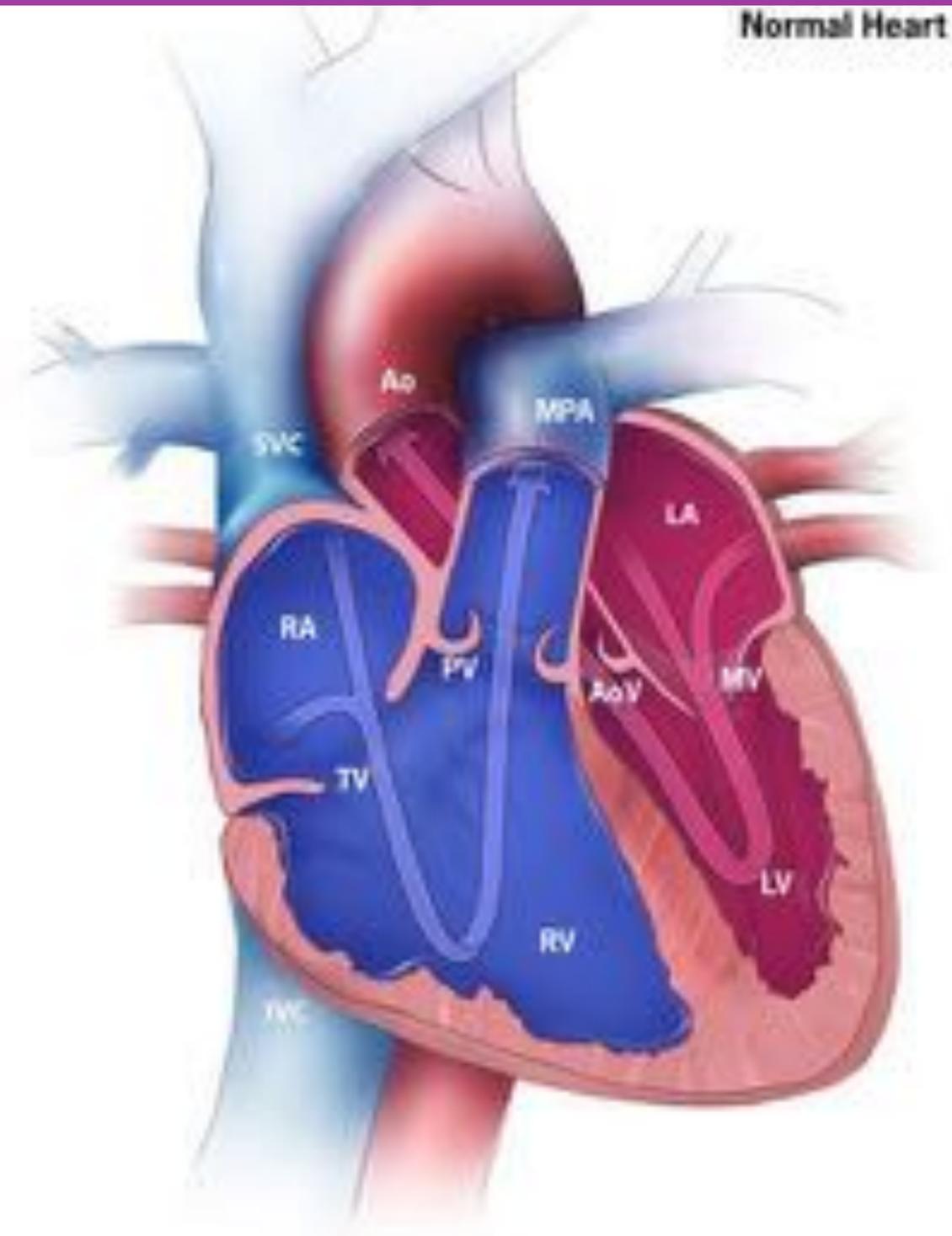
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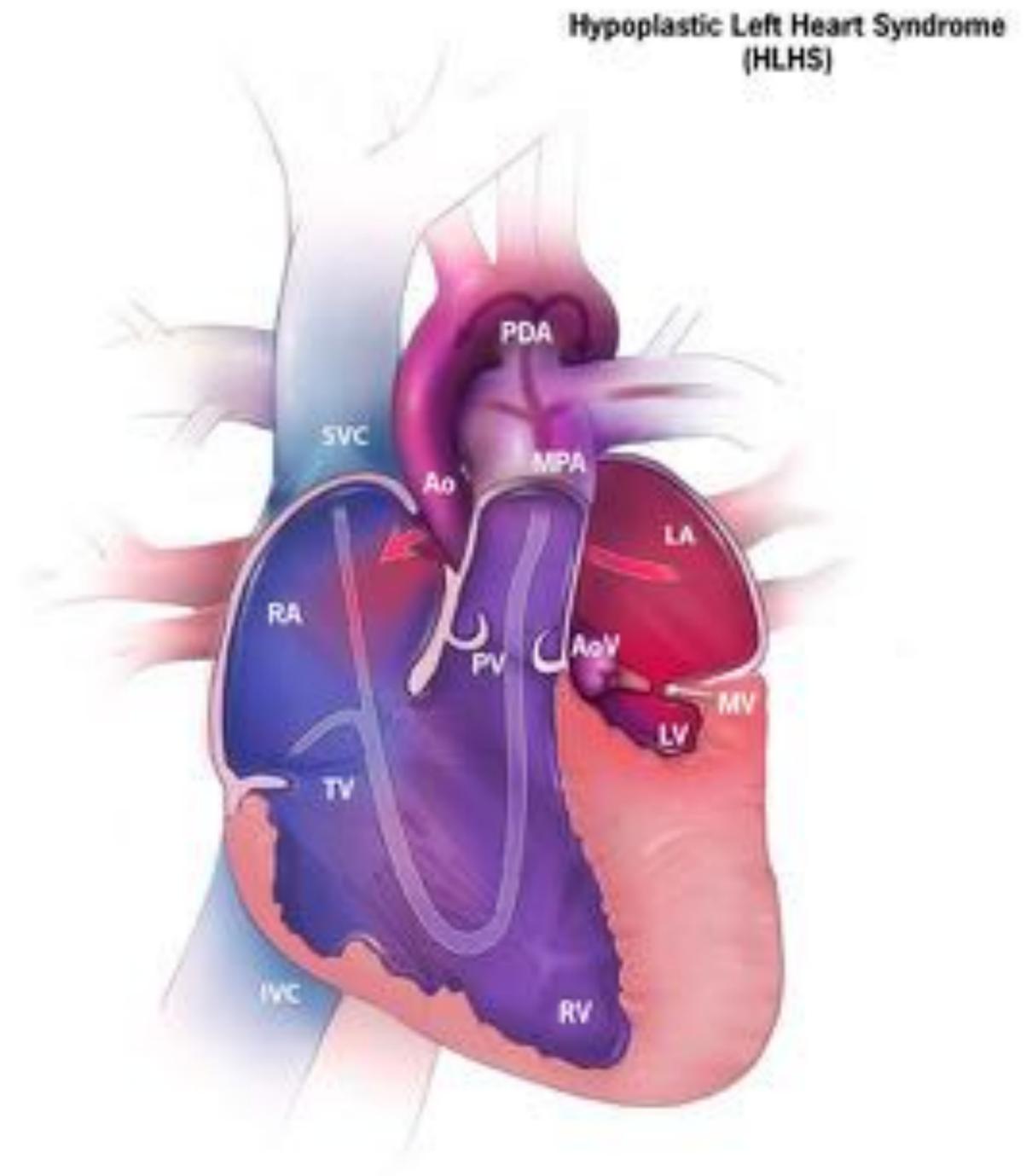
Cardiopathies complexes irréparables



RA, Right Atrium
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LV, Left Ventricle

SVC, Superior Vena Cava
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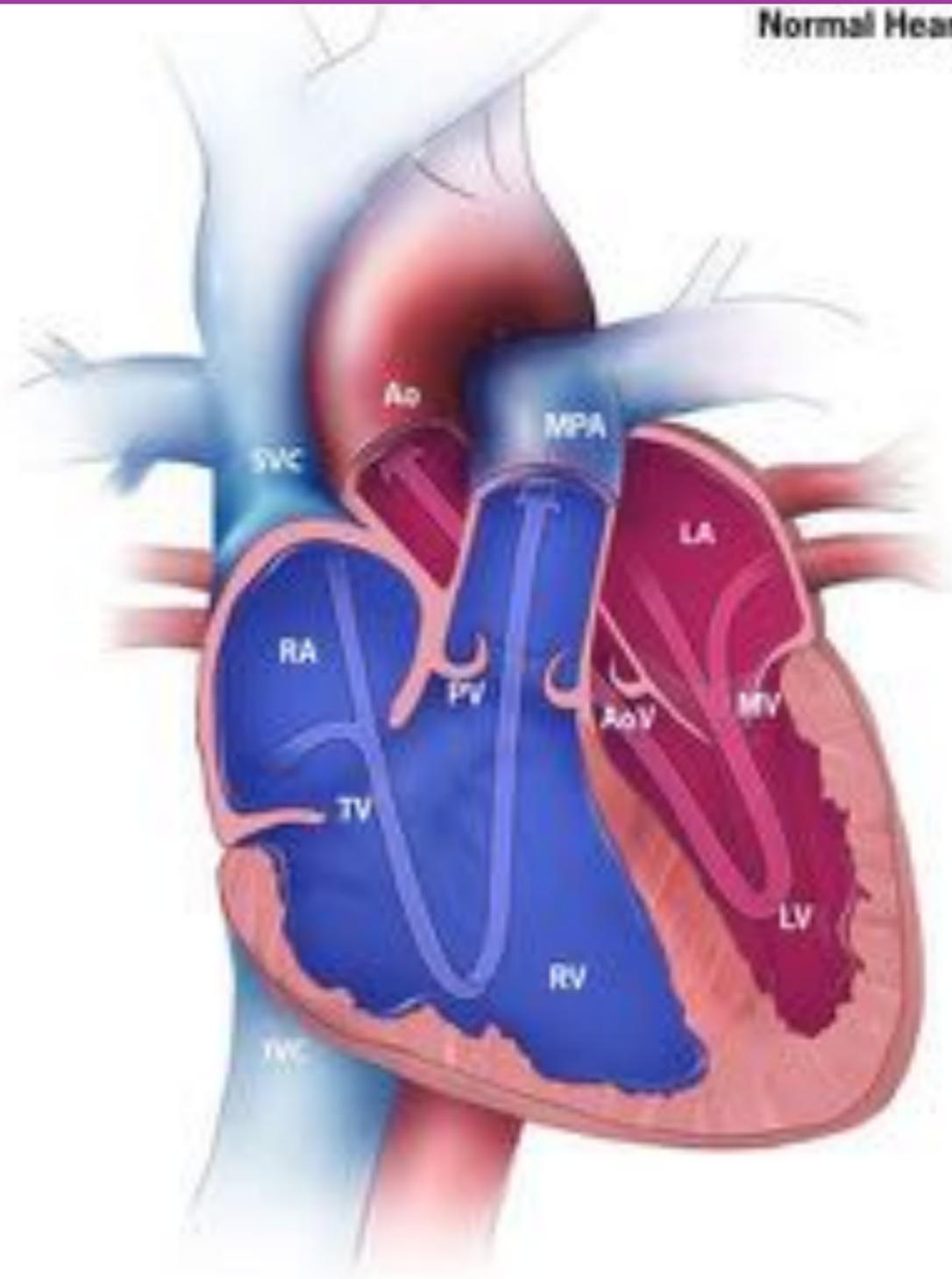
SVC, Superior Vena Cava
IVC, Inferior Vena Cava
MPA, Main Pulmonary Artery
Ao, Aorta
PDA, Patent Ductus Arteriosus

TV, Tricuspid Valve
MV, Mitral Valve
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Cardiopathies complexes irréparables

Normal Heart

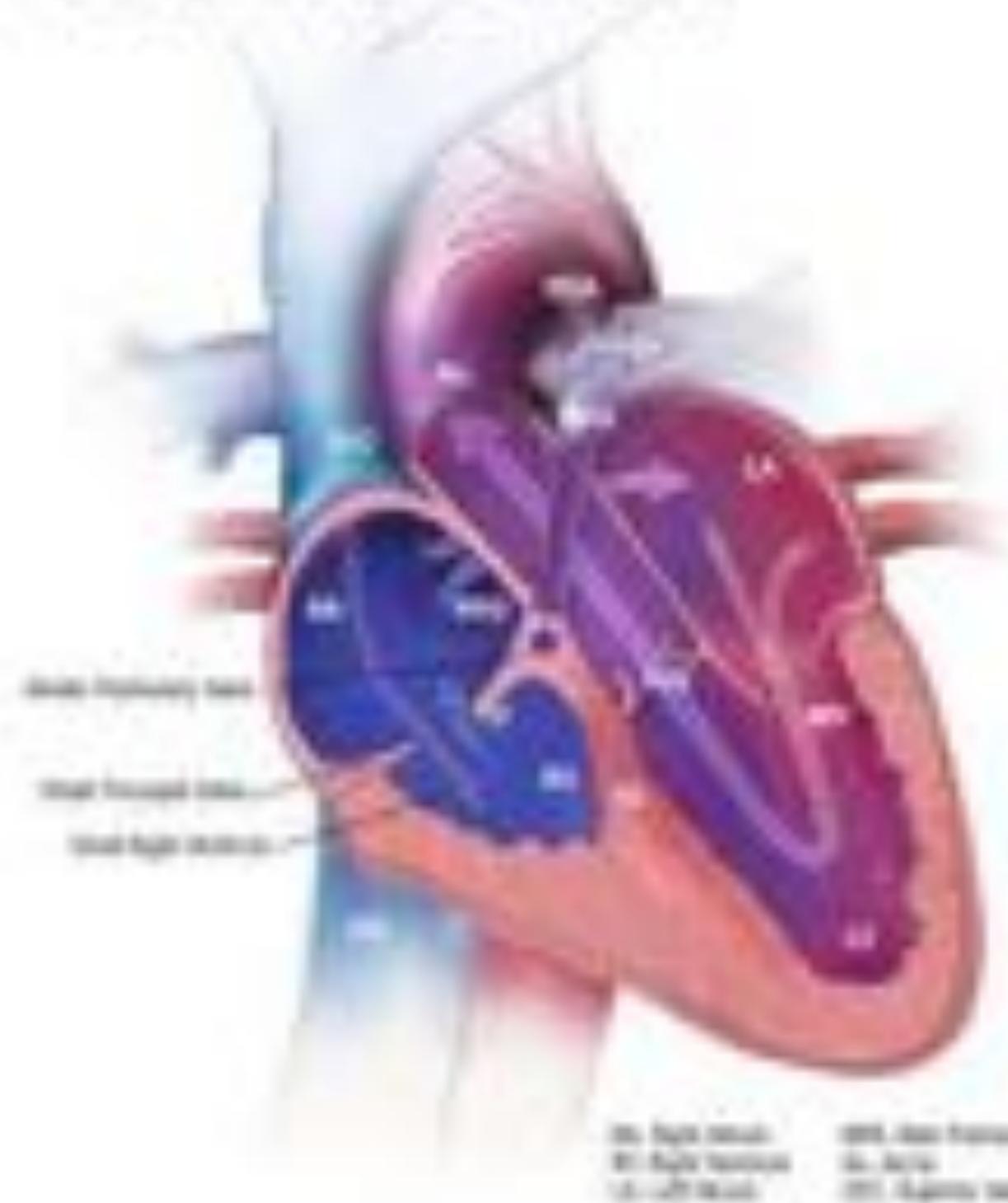


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Pulmonary Stenosis with Atrial Septal Defect

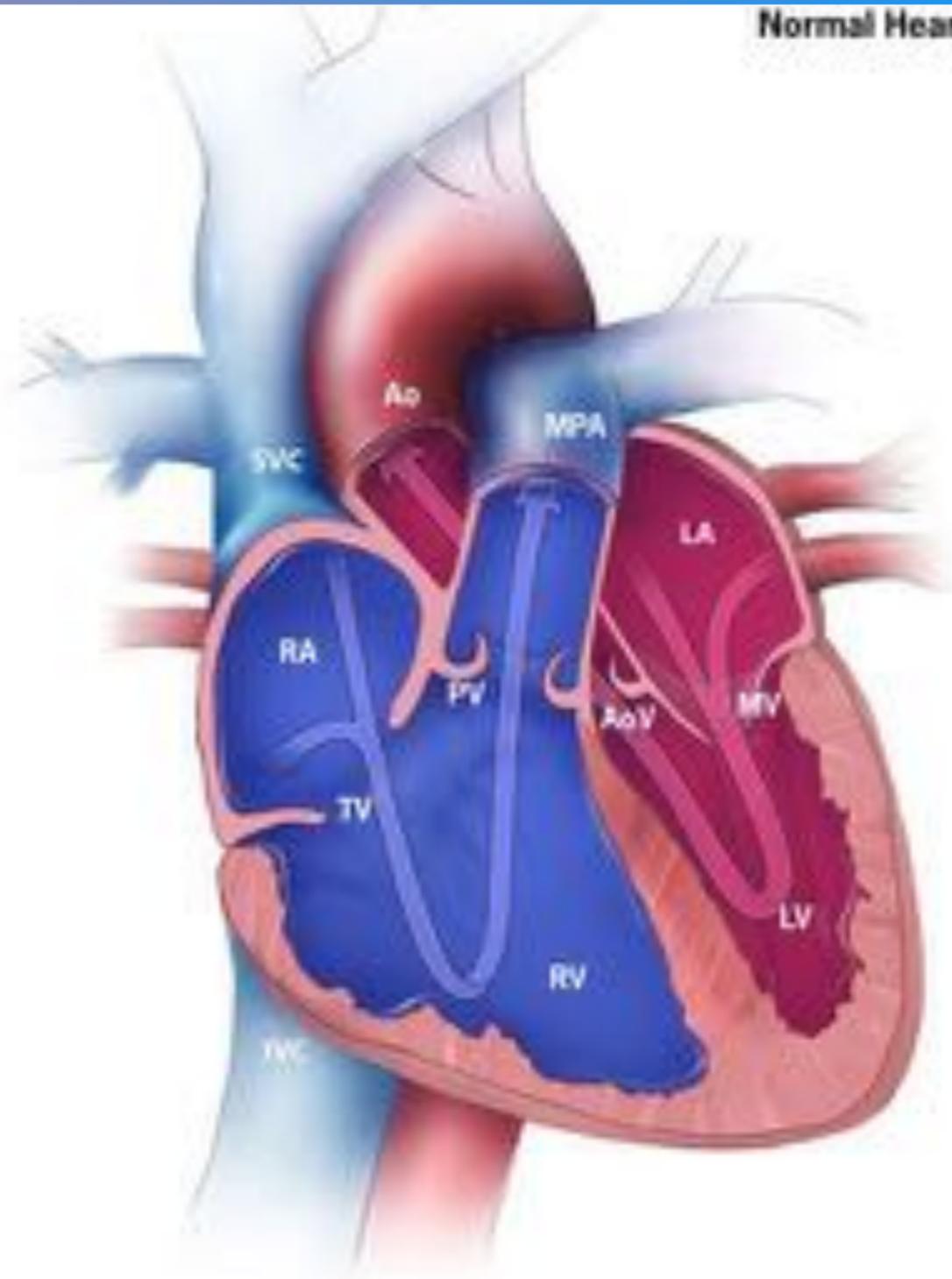


Block pulmonary flow
Flow through ASD
Flow right ventricle

RV Right Ventricle
RA Right Atrium
LA Left Atrium
LV Left Ventricle
SVC Superior Vena Cava
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Cardiopathies complexes réparables

Normal Heart

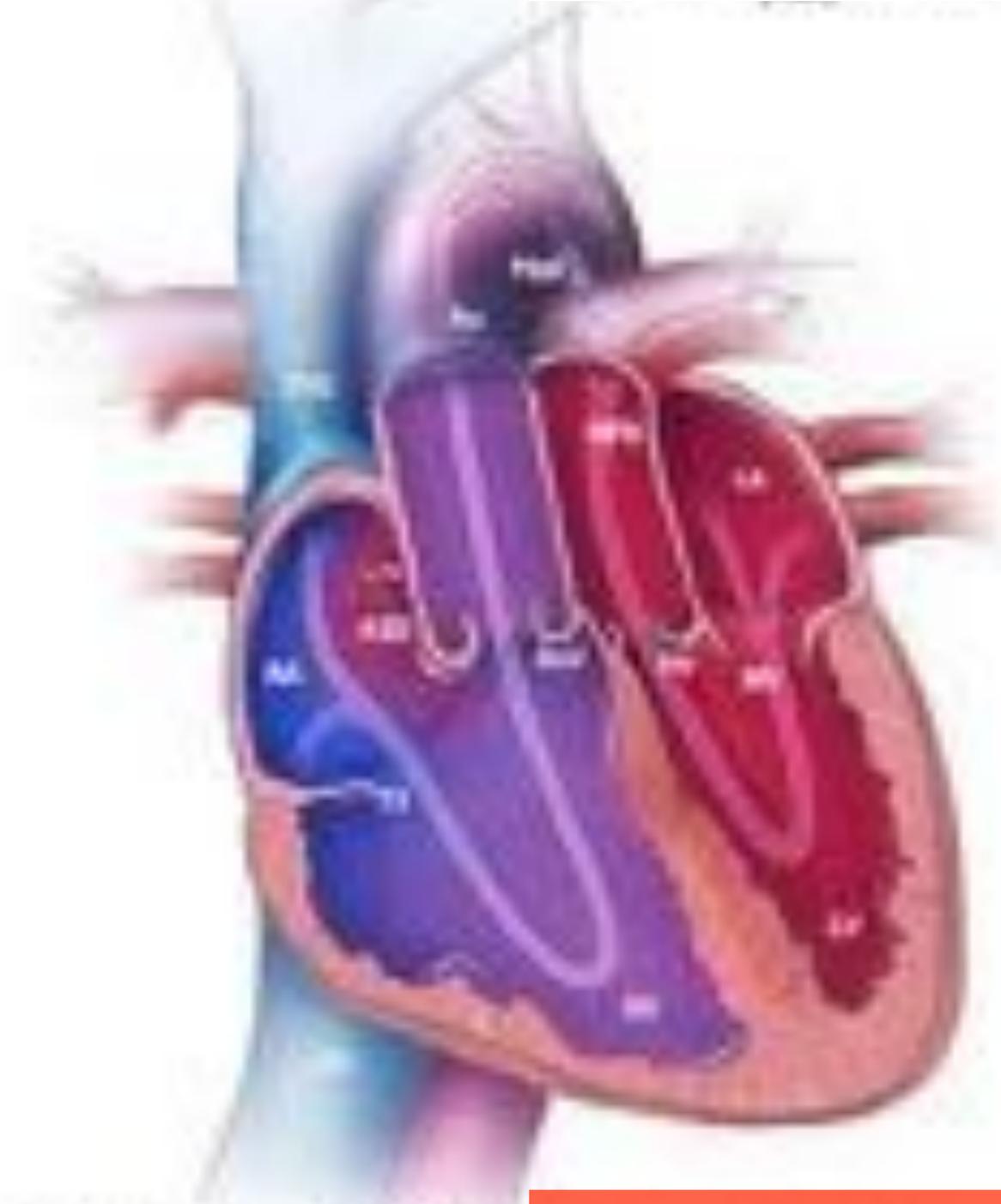


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Transposition of the great arteries
TGA

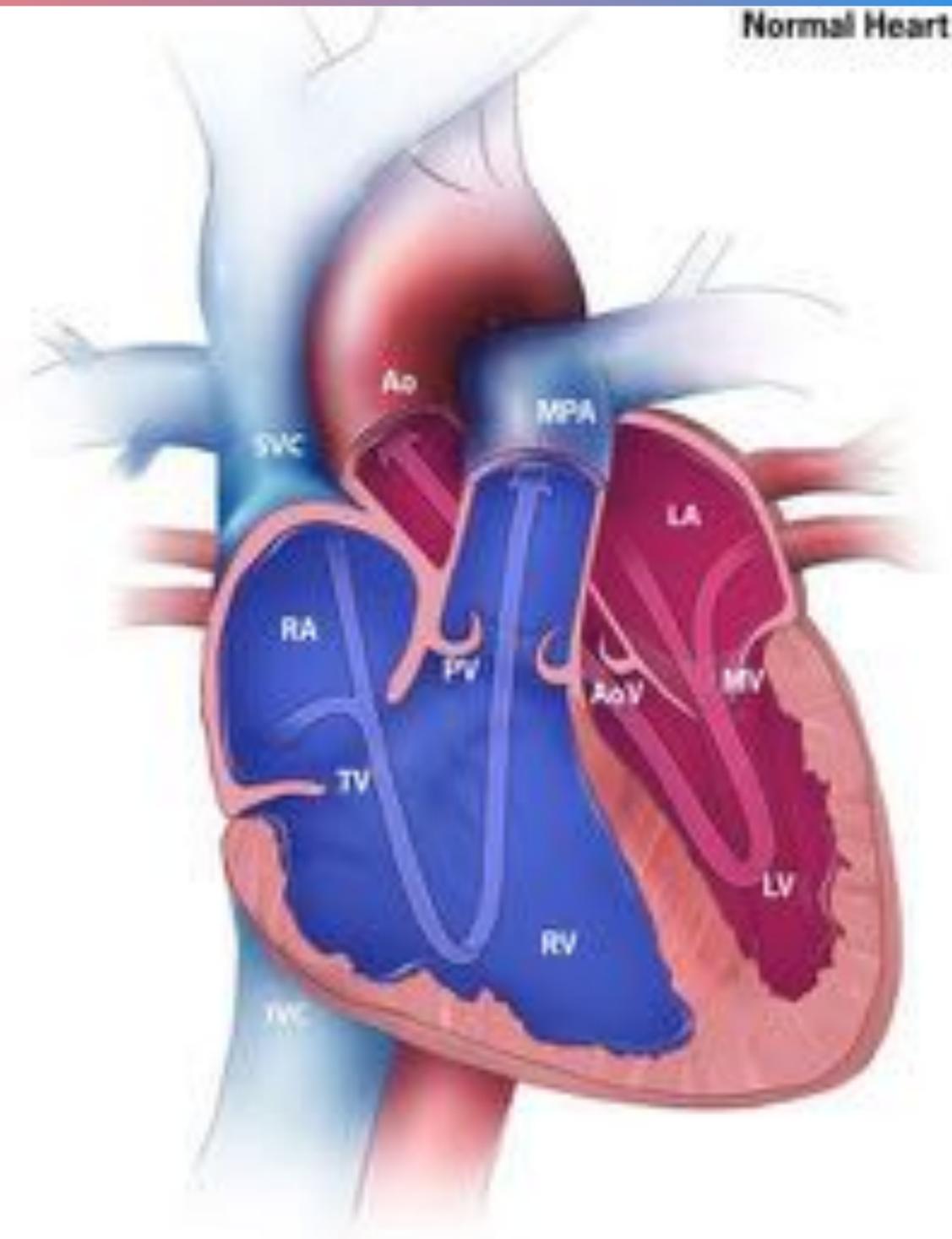


Risque de réintervention faible



M3C

Cardiopathies complexes réparables



Normal Heart

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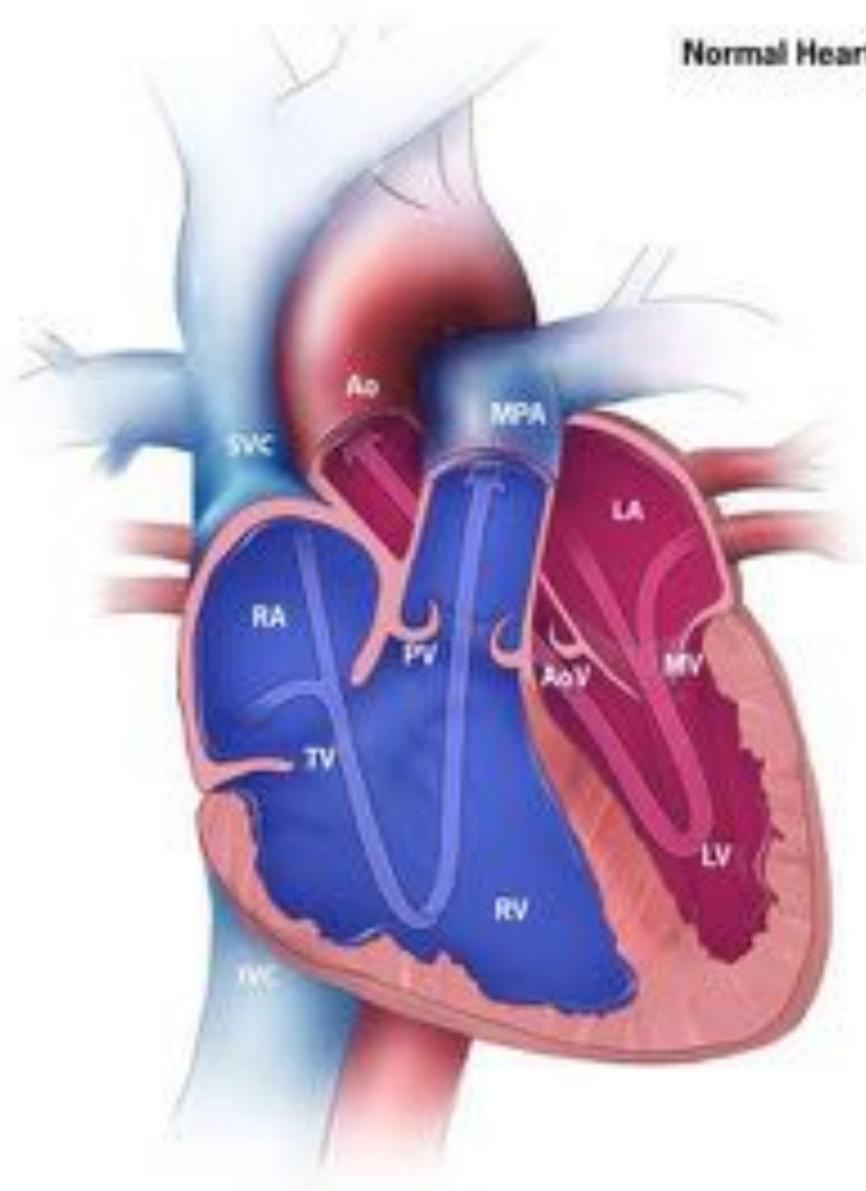
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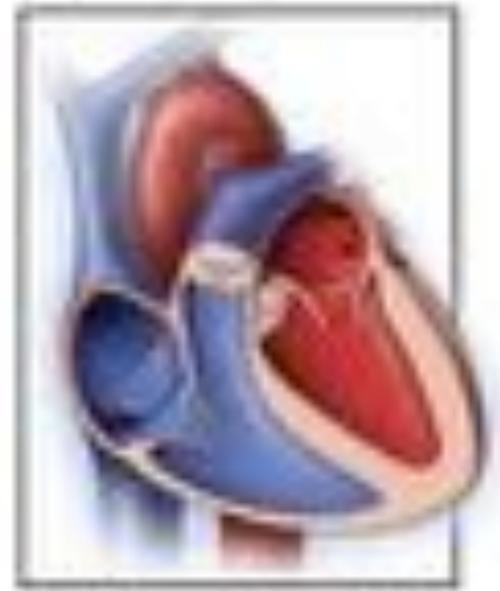
Tetralogy of Fallot
ToF

Risque de réintervention élevé

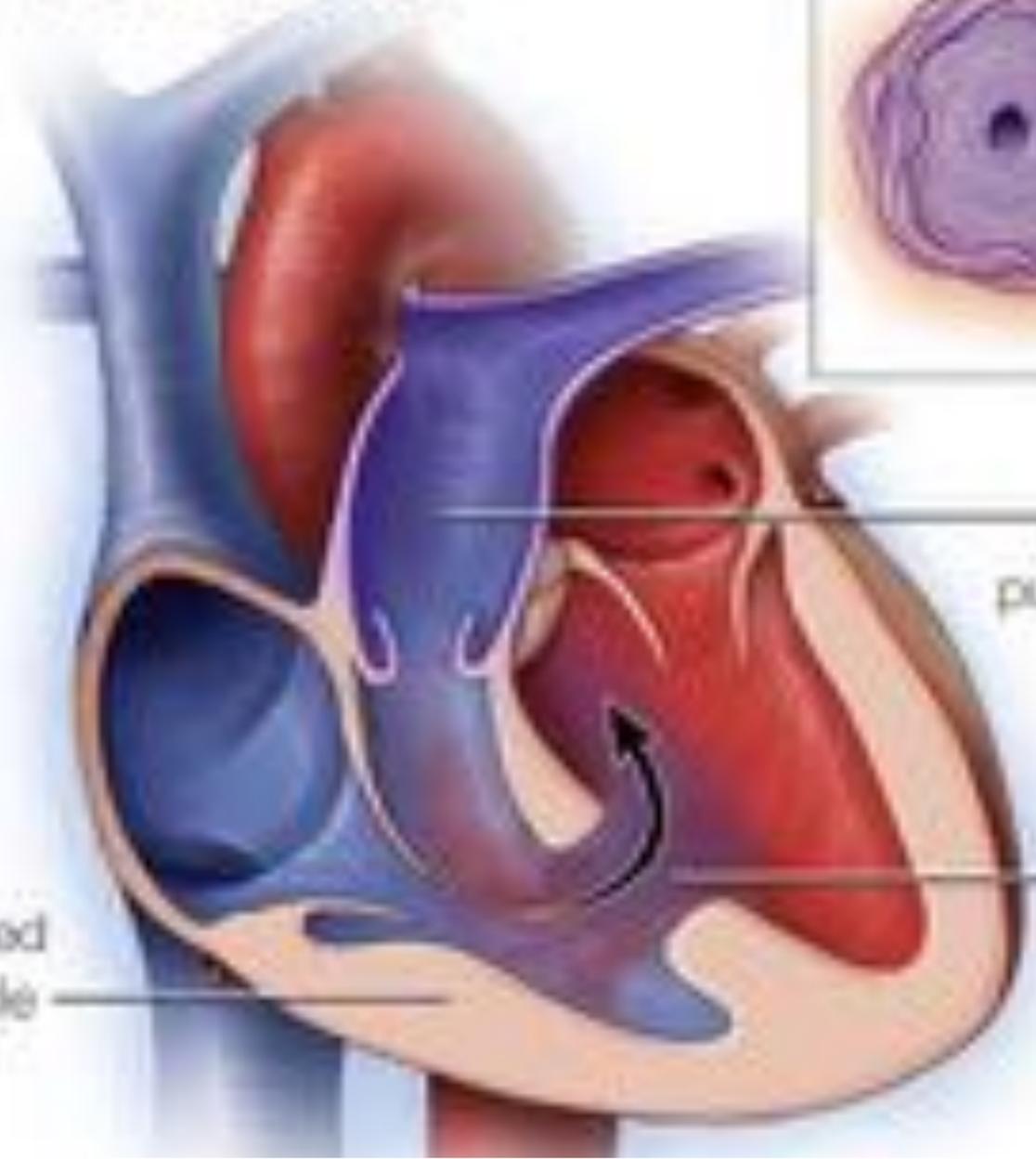


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EISENMENGER SYNDROME

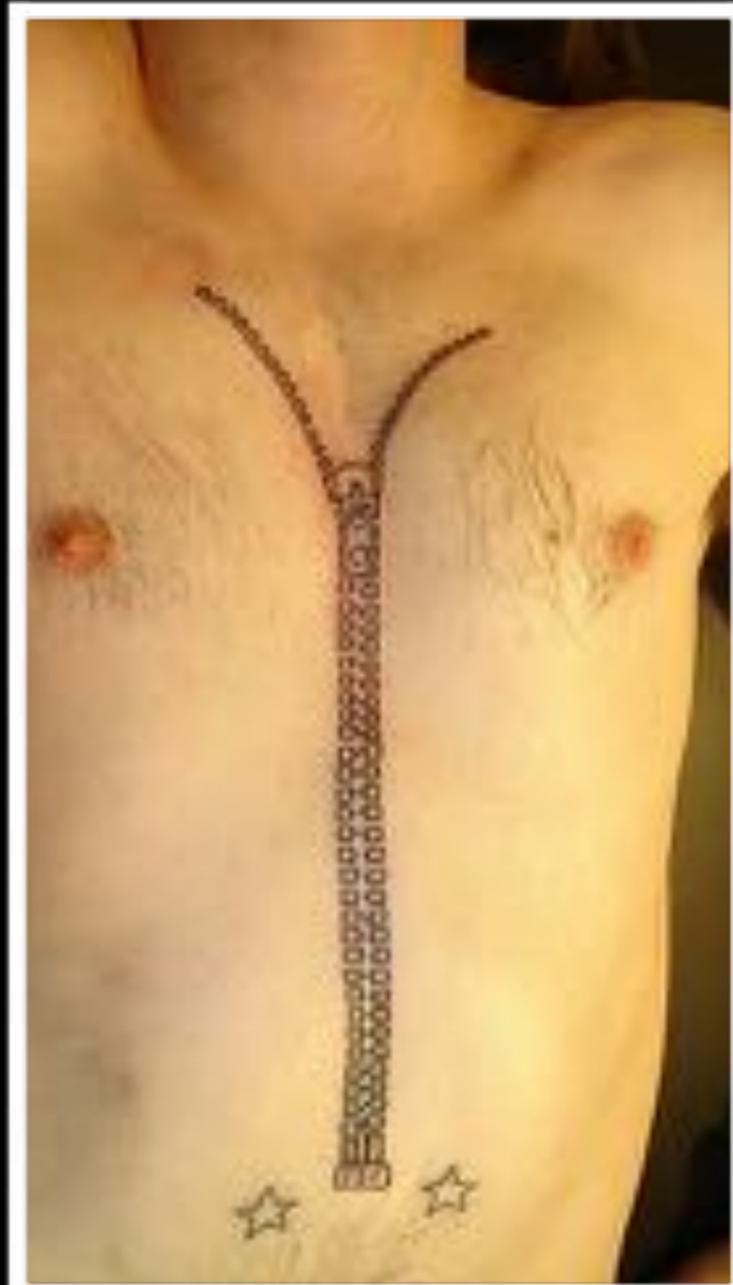


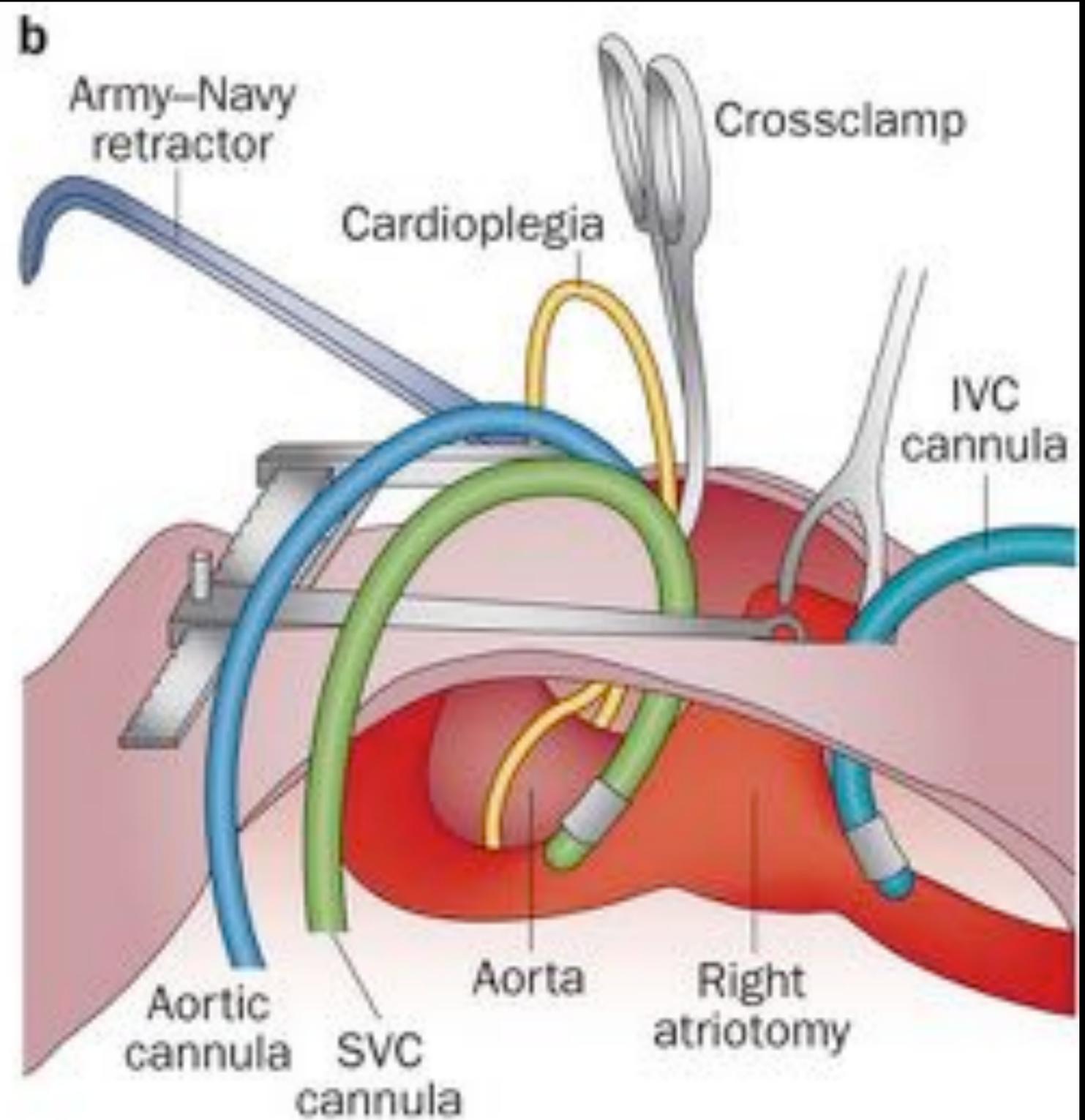
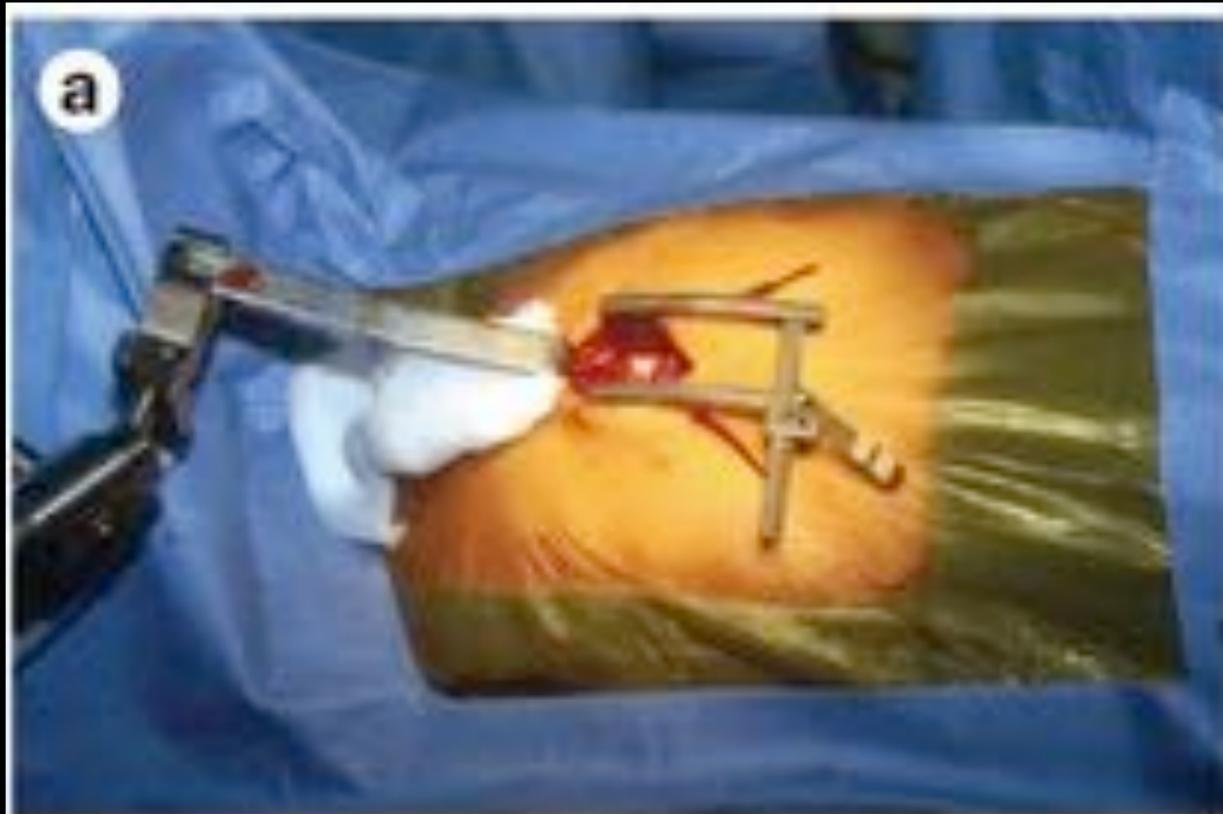
Normal heart

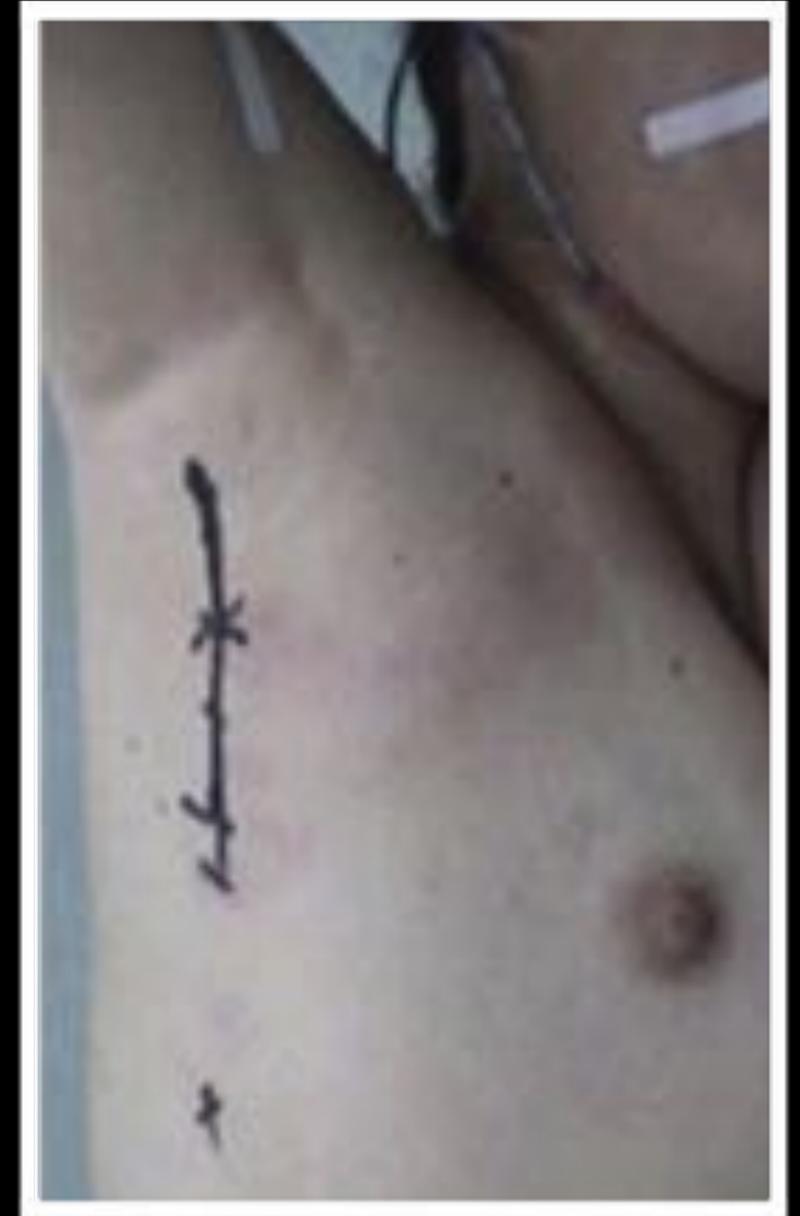
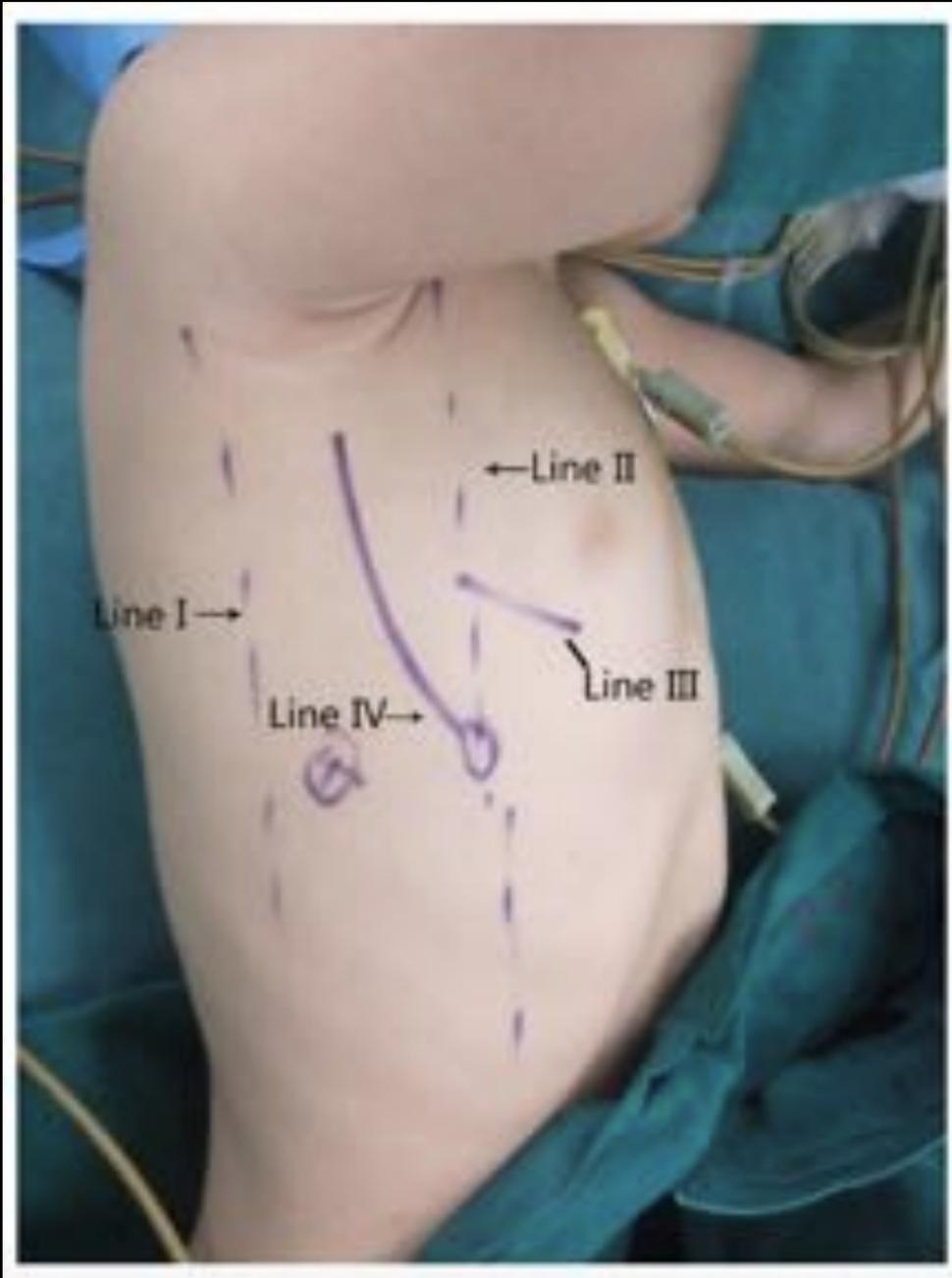


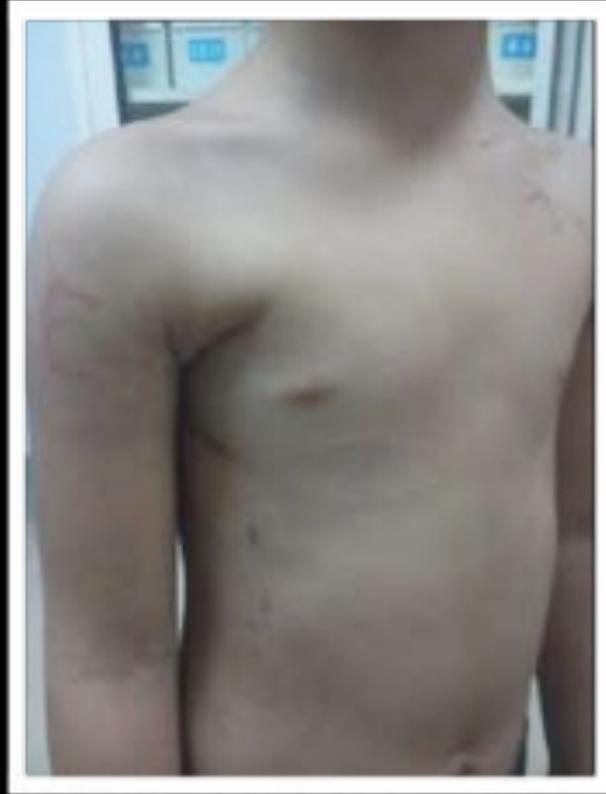
Pulmonary artery wall thickens toward lungs

Y aura-t-il des séquelles ou des complications ?









Pourra-t-il faire du sport ?





Recevra-t-il des médicaments ?

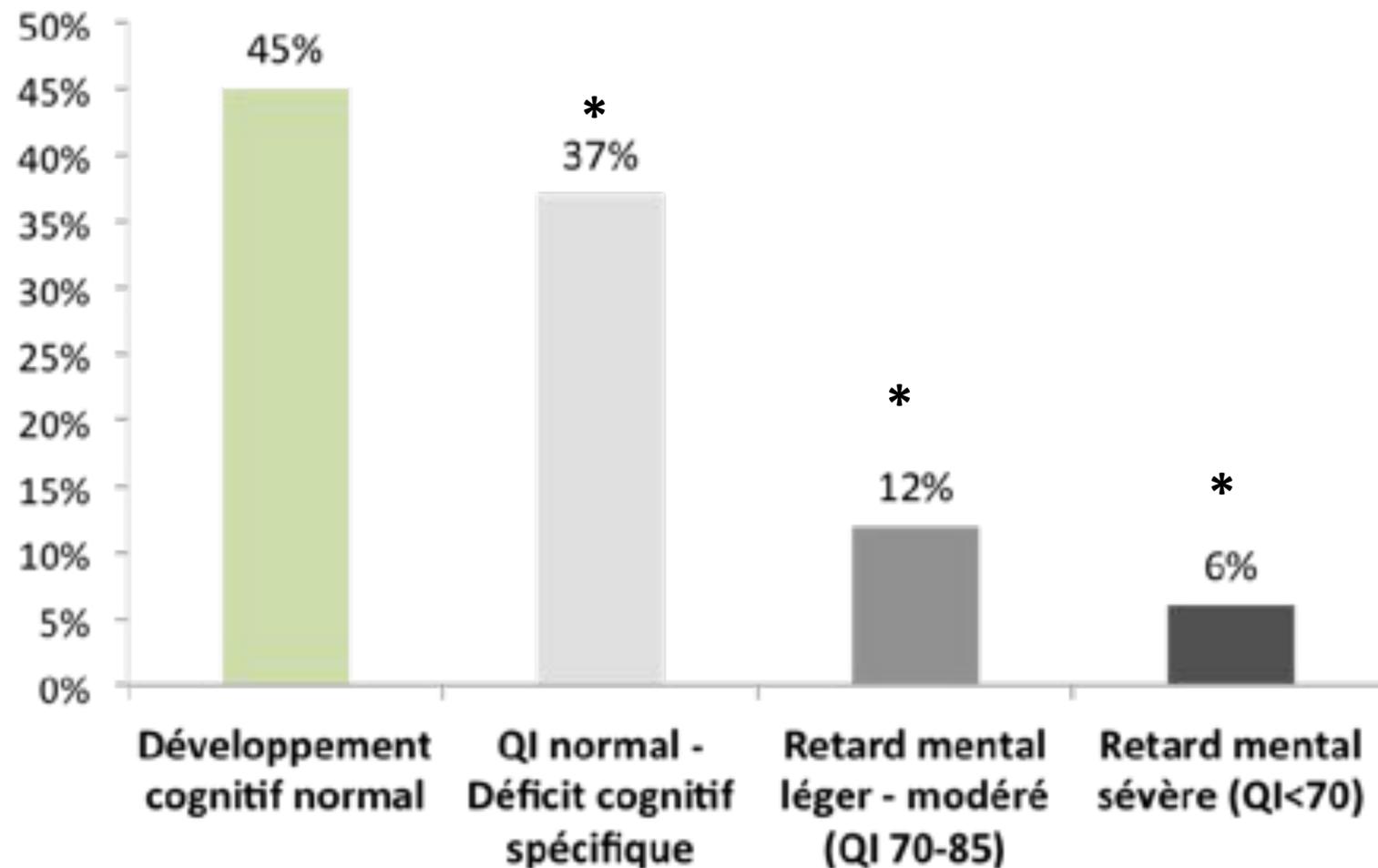
Pourra-t-il avoir des séquelles neurologiques ou intellectuelles ?



Pronostic neuro-développemental des CC

Etude en population EPICARD

- **Etude prospective en population:** tous les cas des CC entre 2005-2008 en IDF
- Examen cognitif complet (test standardisé – K-ABC II) à **3 ans**
- **Résultats sur 415 enfants ayant une CC isolée (CC complexes et mineures)**



Facteurs de risque:

- CC complexes et/ou
- Chirurgie cardiaque réparatrice ou palliative avant l'âge de 1 an
- AG < 39 semaines
- Facteurs socio-démographiques

* p<0.05

Devrons-nous venir souvent en consultation ?



Jusqu'à quel âge ?



1 an

3 ans

10 ans

18 ans

30 ans

60 ans

80 ans

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75015 Paris

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Thank you

