AHA/ACC GUIDELINE

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary

Nouvelles recommandations nord-américaines sur les anomalies coronaires congénitales

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PRACTICE GUIDELINE: FULL TEXT

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Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

JACC
JOURNAL of the AMERICAN COLLEGE of CARDIOLOGY

Warnes CA et al. J Am Coll Cardiol 2008

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ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

EUROPEAN SOCIETY OF CARDIOLOGY®

Baumgartner H et al. Eur Heart J 2010.

Circulation

AHA/ACC GUIDELINE

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary

Stout KK et al. Circulation 2019.

DEFINITIONS

- Cardiopathie congénitale :

  anomalie toujours présente à la naissance

- Cardiopathie génétique :

  anomalie pas forcément présente à la naissance
  anomalie d'un gène ou d'un chromosome

- Cardiopathie héréditaire :

  anomalie pas forcément présent à la naissance
  anomalie d'un gène ou d'un chromosome
  transmission parentale

1.4. Scope of the Guideline

The 2018 ACHD guideline is a full revision of the "2008 ACC/AHA Guidelines for the Management of Adults with Congenital Heart Disease,"[S1.4-1] which was the first US guideline to be published on the topic. This revision uses the 2008 ACHD guideline as a framework and incorporates new data and growing ACHD expertise to develop recommendations. Congenital heart disease (CHD) encompasses a range of structural cardiac abnormalities present before birth attributable to abnormal fetal cardiac development but does not include inherited disorders that may have cardiac manifestations such as Marfan syndrome or hypertrophic cardiomyopathy. Also not included are anatomic variants such as patent foramen ovale. Valvular heart disease (VHD) may be congenital, so management overlaps with the "2014 AHA/ACC Guidelines for the Management of Patients With Valvular Heart Disease,"[S1.4-2] particularly for bicuspid aortic valve (BAV) disease. Where overlap exists, this document focuses on the diagnosis and treatment of congenital valve disease when it differs from acquired valve disease, whether because of anatomic differences, presence of concomitant lesions, or differences to consider given the relatively young age of patients with ACHD. This guideline is not intended to apply to children (<18 years of age) with CHD or adults with acquired VHD, heart failure (HF), or other cardiovascular disease.

Marfan

CMH

FOP

Bicuspidie

Fistule coronaire

2018 AHA/ACC Guideline

CHD Anatomy*

II: Moderate Complexity
Repaired or unrepaired conditions
Aorto-left ventricular fistula
Anomalous pulmonary venous connection, partial or total
Anomalous coronary artery arising from the pulmonary artery
Anomalous aortic origin of a coronary artery from the opposite sinus
AVSD (partial or complete, including primum ASD)
Congenital aortic valve disease
Congenital mitral valve disease
Coarctation of the aorta
Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)
Infundibular right ventricular outflow obstruction
Ostium primum ASD
Moderate and large unrepaired secundum ASD
Moderate and large persistently patent ductus arteriosus
Pulmonary valve regurgitation (moderate or greater)
Pulmonary valve stenosis (moderate or greater)
Peripheral pulmonary stenosis
Sinus of Valsalva fistula/aneurysm
Sinus venosus defect
Subvalvar aortic stenosis (excluding HCM; HCM not addressed in these guidelines)
Supravalvar aortic stenosis
Straddling atrioventricular valve
Repaired tetralogy of Fallot
VSD with associated abnormality and/or moderate or greater shunt

Stout KK et al. Circulation 2019.

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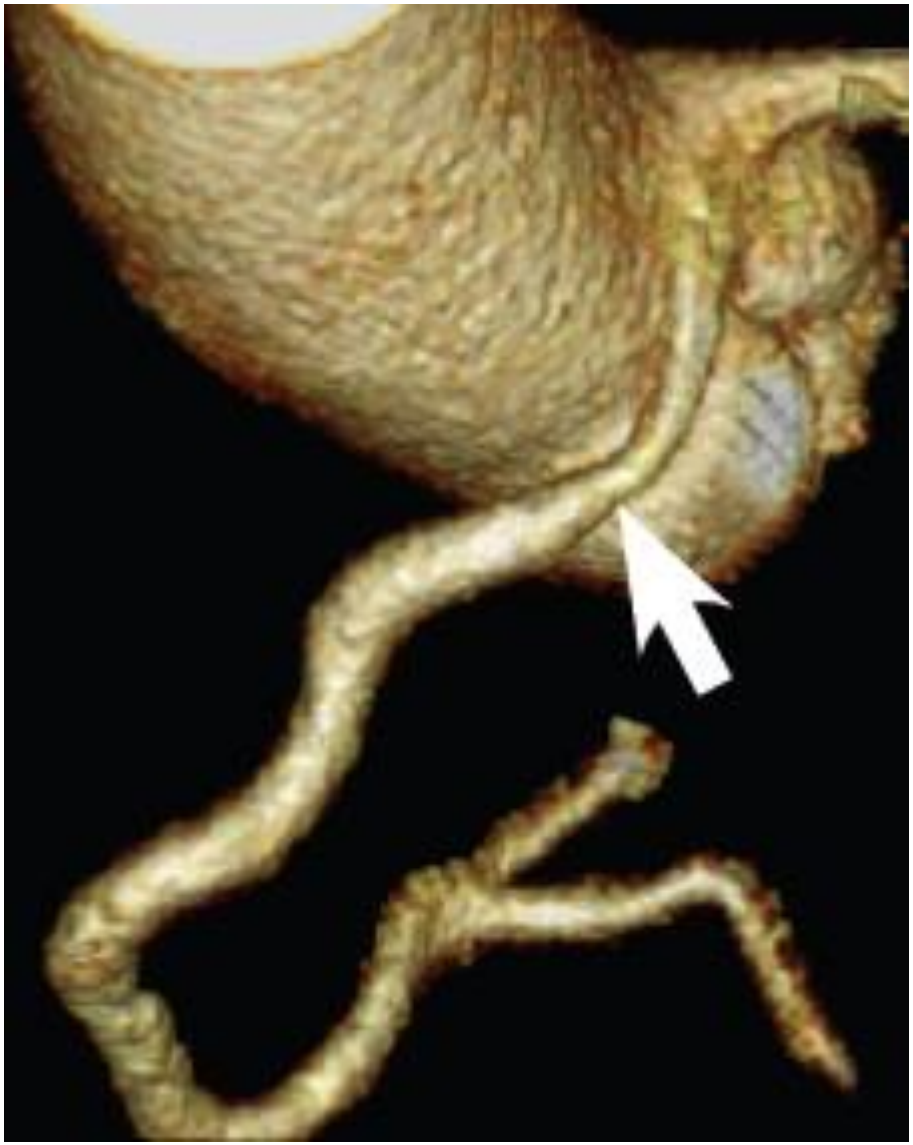
Activité interventionnelle percutanée 2018

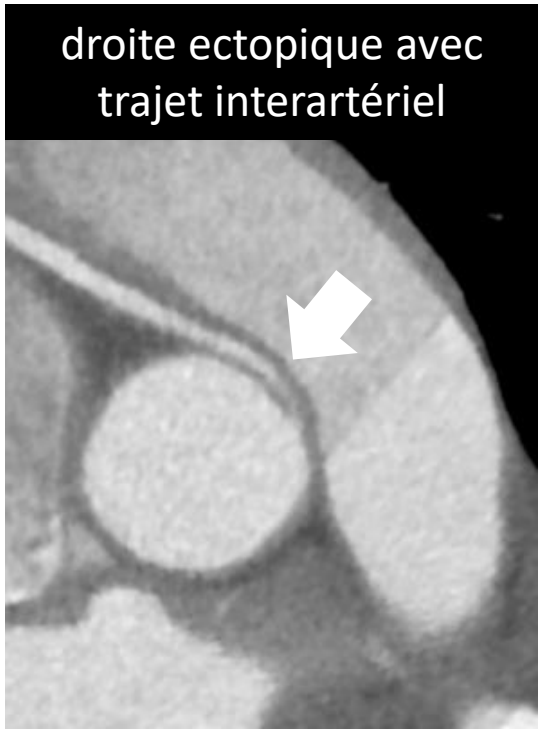
Actes	Nombre	%
Angioplasties coronaires	920	
Procédures valvulaires aortiques	222	
Fermetures FOP	107	
Procédures valvulaires mitrales	84	
Fermetures auricule gauche	25	
Fermetures CIA	22	1.6
Fermetures fuites paravalvulaires	11	
Corrections anomalies coronaires	3	0.2
Total	**1396**	

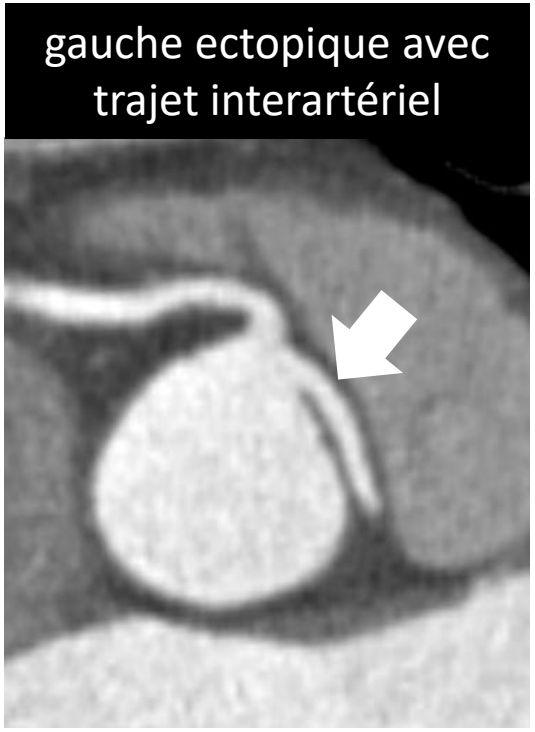
ANOMALOUS CORONARY CONNECTIONS



CONNEXIONS CORONAIRES ANORMALES

Formes anatomiques à risque

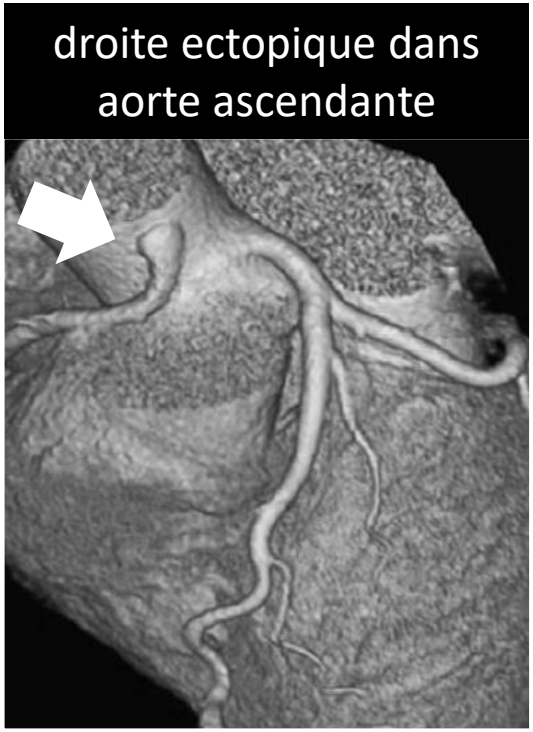


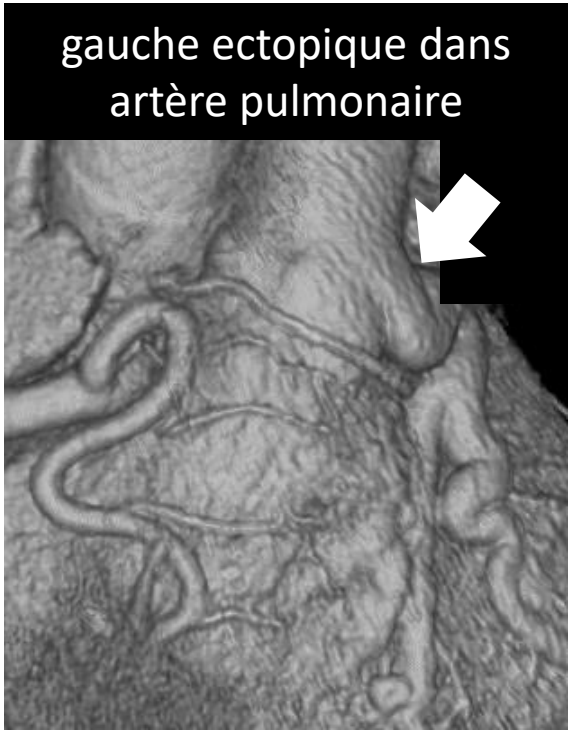


Risques connus pour : ischémie myocardique, troubles rythmiques ventriculaires (tachycardie, fibrillation) et mort subite.

CONNEXIONS CORONAIRES ANORMALES

Formes anatomiques à risque





Risques connus pour : ischémie myocardique, troubles rythmiques ventriculaires (tachycardie, fibrillation) et mort subite.

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Warnes CA et al. J Am Coll Cardiol 2008

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8.5. Recommendations for Congenital Coronary Anomalies of Ectopic Arterial Origin

Class I

3. Surgical coronary revascularization should be performed in patients with any of the following indications:
   a. Anomalous left main coronary artery coursing between the aorta and pulmonary artery. (*Level of Evidence: B*)
   b. Documented coronary ischemia due to coronary compression (when coursing between the great arteries or in intramural fashion). (*Level of Evidence: B*)
   c. Anomalous origin of the right coronary artery between aorta and pulmonary artery with evidence of ischemia. (*Level of Evidence: B*)

Warnes CA et al. J Am Coll Cardiol 2008

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The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric Cardiology (AEPC)

- Atrial septal defect
- Ventricular septal defect
- Atrioseptal defect
- Patent ductus arteriosus
- Left ventricular outflow tract obstruction
- Coarctation of the aorta
- Marfan syndrome
- Right ventricular outflow tract obstruction
- Ebstein's anomaly
- Tetralogy of Fallot
- Pulmonary atresia with ventricular septal defect
- Transposition of the great arteries
- Univentricular heart
- Congenitally corrected transposition of the great arteries
- Eisenmenger syndrome and severe pulmonary arterial hypertension

ESC GUIDELINES

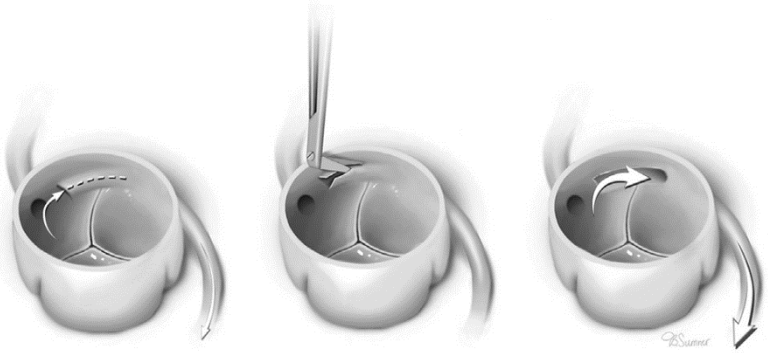
Eur Heart J 2010

anomalies coronaires congénitales : **non citées**

CORRECTION CHIRURGICALE

CHIRURGIE

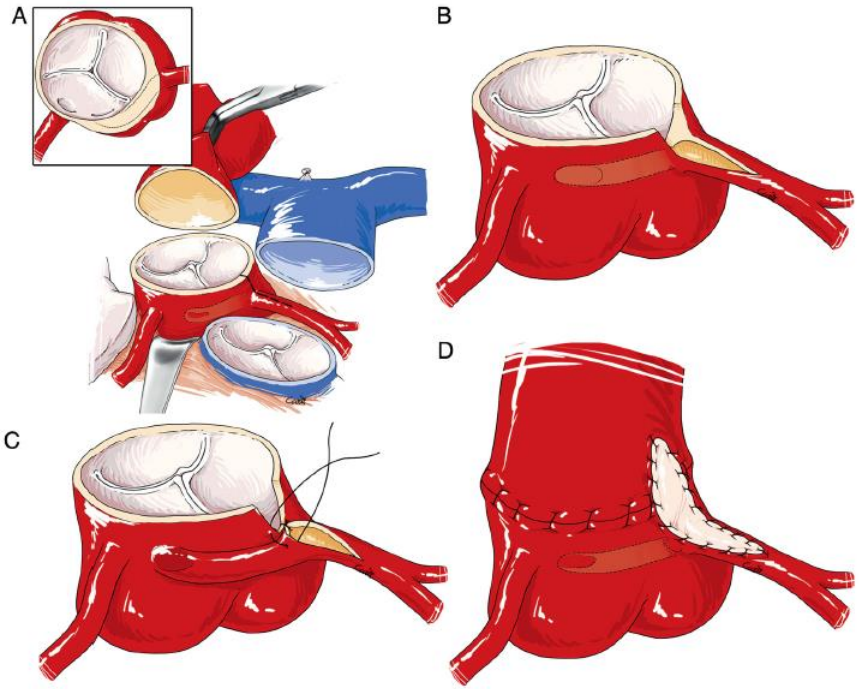
Exérèse bandelette aortique avec néo-ostium (*unroofing*)



Mery CM. *Semin Thoracic Surg* 2014

CHIRURGIE

Néo-ostium avec plastie



Gaudin R. *Multimed Man Cardiothorac Surg* 2014

CONNEXIONS CORONAIRES ANORMALES

MORT SUBITE

- Estimation difficile du risque individuel de mort subite.
- Absence de score pour stratifier le risque.
- Risque ANOCOR gauche très supérieur/ANOCOR droite (rapport de 20 à 1).

risque annuel de mort subite (estimation)
population 15-35 ans sportive

0.2% pour ANOCOR gauche

0.01% pour ANOCOR droite

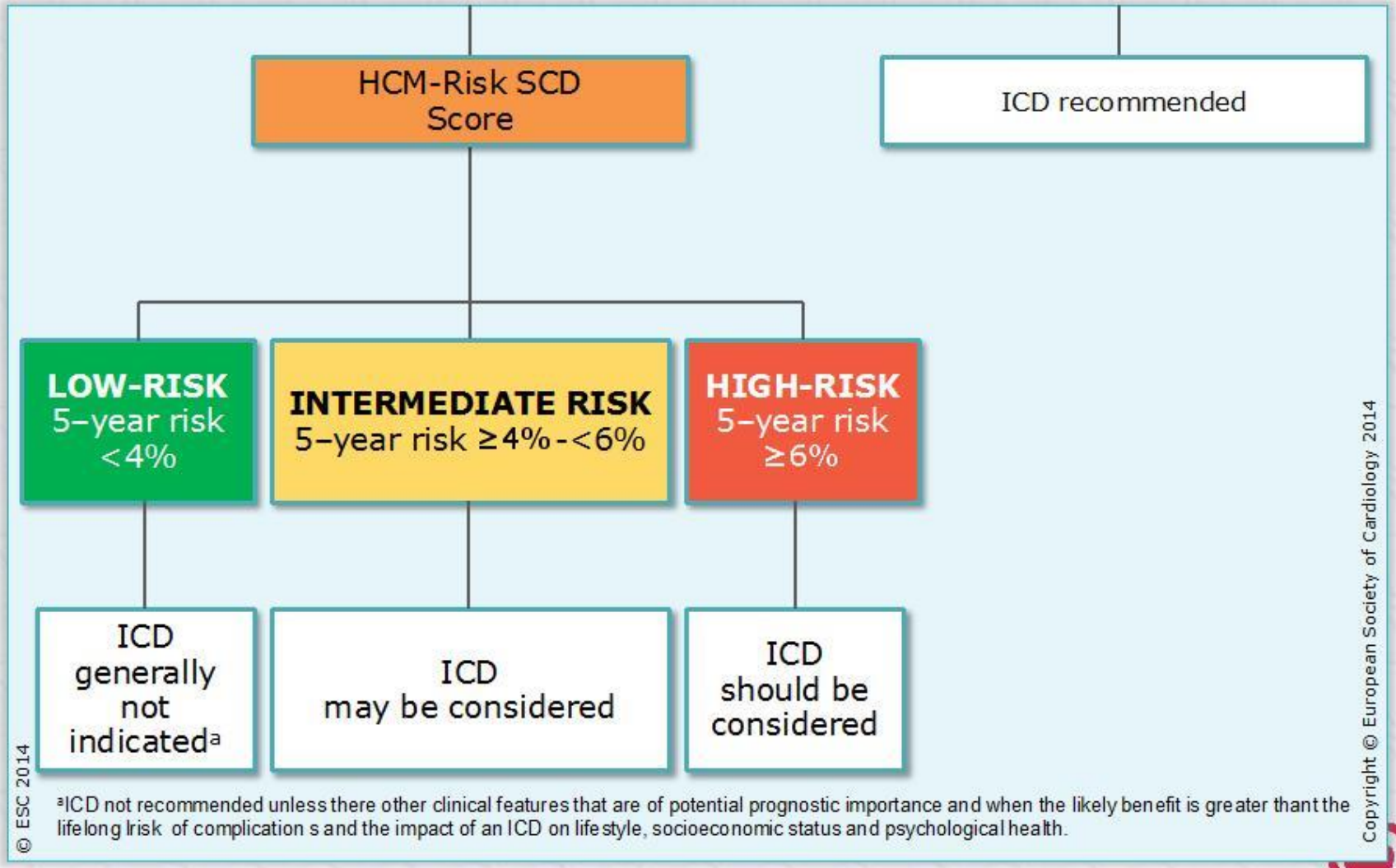
Mery CM. *Semin Thoracic Surg* 2014.

MORT SUBITE et CARDIOPATHIES non ACQUISES (estimations)

Cardiopathie	Incidence mort subite (cas annuels pour 1.000 individus)
Tachycardie ventriculaire catécholergique	**15**
Cardiomyopathie hypertrophique	**10-20**
Syndrome de Brugada	**10**
Syndrome du QT long	**5-10**
Cardiomyopathie dilatée idiopathique	**5-10**
Dysplasie arythmogène du ventricule droit	**5-10**
Anomalie de connexion coronaire gauche	**2**
Syndrome de pré-excitation ventriculaire	**1**
Anomalie de connexion coronaire droite	**0.1**

Aubry P et al. Ann Cardiol Angéiol 2017.

Flow chart for ICD implantation



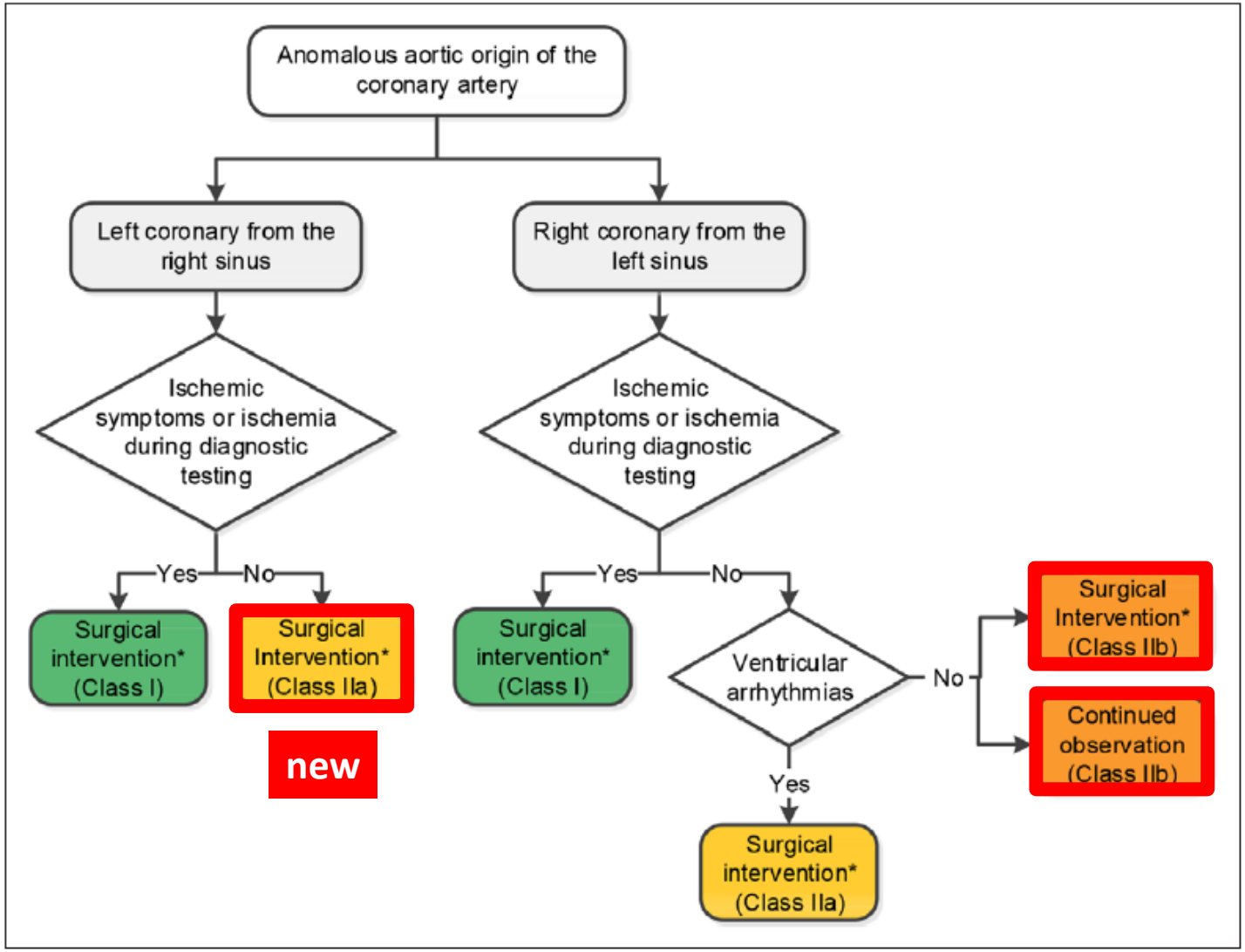
European Heart Journal (2014);35:2733–2779 - doi:10.1093/eurheartj/ehu284

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary

COR	LOE	Recommendations
Therapeutic		
I	B-NR	1. Surgery is recommended for AAOCA from the left sinus or AAOCA from the right sinus for symptoms or diagnostic evidence consistent with coronary ischemia attributable to the anomalous coronary artery.[S4.4.5.2-1–S4.4.5.2-3]
IIa	C-LD	2. Surgery is reasonable for anomalous aortic origin of the left coronary artery from the right sinus in the absence of symptoms or ischemia.[S4.4.5.2-4–S4.4.5.2-6]
IIa	C-EO	3. Surgery for AAOCA is reasonable in the setting of ventricular arrhythmias.
IIb	B-NR	4. Surgery or continued observation may be reasonable for asymptomatic patients with an anomalous left coronary artery arising from the right sinus or right coronary artery arising from the left sinus without ischemia or anatomic or physiological evaluation suggesting potential for compromise of coronary perfusion (eg, intramural course, fish-mouth-shaped orifice, acute angle).[S4.4.5.2-4–S4.4.5.2-6]

Stout KK et al. Circulation 2019

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary



5. Anomalous aortic origin of the coronary artery.
cal intervention to involve unroofing or coronary revascularization for patients with concomitant fixed obstruction.

Stout KK et al. Circulation 2019

Anomalous Aortic Origin of a Coronary Artery is Always a Surgical Disease

Pascal R. Vouhé

Anomalous aortic origin of a coronary artery is a congenital anomaly in which a major coronary artery arises from the wrong sinus of Valsalva (left coronary from right sinus or right coronary from left sinus) and courses between the great arteries before reaching its normal epicardial course. Although the risk of sudden death is clearly established, the indications for surgery remain controversial. The risk of sudden death is increased in symptomatic patients, in anomalous left coronary artery, as well as in the presence of some risk factors (intense physical activity, young age [<35 years], aggravating anatomical features [intramural interarterial course, slit-like ostium, acute angle of take-off]). As far as is currently known, surgical management using an anatomical repair can prevent sudden death, provided that normal coronary anatomy and function are achieved and that extensive return-to-play testing is performed. A precise evaluation of the benefit/risk ratio is mandatory on an individual basis, but surgery may be indicated in the vast majority of patients.
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 19:25-29

Activité chirurgicale 2017

Base Nationale Publique et Privée - 2018 (mise à jour hebdomadaire)

Répartition des GHM pour l'acte CCAM

DDEA001 : Réimplantation d'une artère coronaire pour anomalie congénitale d'origine, par thoracotomie avec CEC

CMD	GHM	Libellé	Effectif	DMS
05	05C062	Autres interventions cardiothoraciques, âge supérieur à 1 an, ou vasculaires quel que soit l'âge, avec circulation extracorporelle, niveau 2	29	8,97
05	05C061	Autres interventions cardiothoraciques, âge supérieur à 1 an, ou vasculaires quel que soit l'âge, avec circulation extracorporelle, niveau 1	19	8,89
05	05C074	Autres interventions cardiothoraciques, âge inférieur à 2 ans, avec circulation extracorporelle, niveau 4	9	36,11
05	05C064	Autres interventions cardiothoraciques, âge supérieur à 1 an, ou vasculaires quel que soit l'âge, avec circulation extracorporelle, niveau 4	6	33,83
05	05C073	Autres interventions cardiothoraciques, âge inférieur à 2 ans, avec circulation extracorporelle, niveau 3	4	18,25
05	05C063	Autres interventions cardiothoraciques, âge supérieur à 1 an, ou vasculaires quel que soit l'âge, avec circulation extracorporelle, niveau 3	4	18,00
05	05C072	Autres interventions cardiothoraciques, âge inférieur à 2 ans, avec circulation extracorporelle, niveau 2	1	15,00
05	05C071	Autres interventions cardiothoraciques, âge inférieur à 2 ans, avec circulation extracorporelle, niveau 1	1	7,00
			73	

PREVALENCE des CARDIOPATHIES non ACQUISES à RISQUE de MORT SUBITE (estimations)

Cardiopathie	Prévalence de la cardiopathie (nombre de cas pour 100,000 individus)
Anomalies de connexion des artères coronaires[a]	340 (300/40)
Cardiomyopathie hypertrophique[b]	200
Syndrome de pré-excitation ventriculaire	150
Syndrome du QT long	50
Cardiomyopathie dilatée[c]	40
Dysplasie arythmogène ventriculaire droite	40
Syndrome de Brugada	20
Tachycardie ventriculaire catécholergique	10

[a] Formes anatomiques reconnues à risque, prévalences et incidences globales et respectives des ANOCOR droites et gauches.
[b] Non liées à une pathologie entraînant une surcharge ventriculaire gauche de volume ou de pression.
[c] Non liées à une cardiopathie ischémique ou à une valvulopathie.
[d] Formes congénitales/génétiques de l'enfant.

Aubry P et al. Ann Cardiol Angéiol 2017.

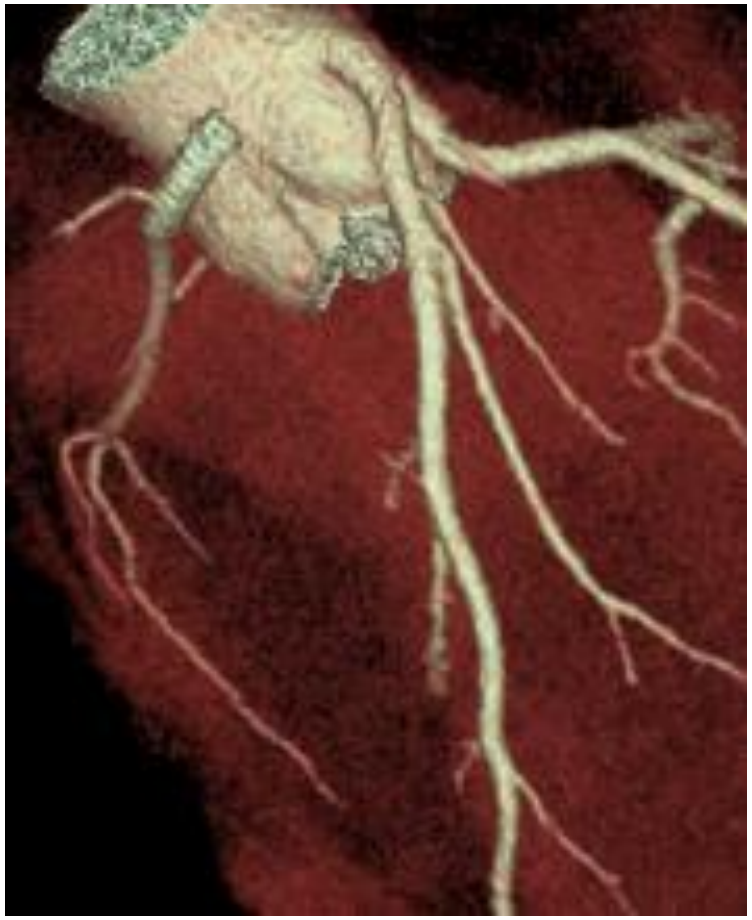
SURGICAL CORRECTION of ANOMALOUS CORONAY CONNECTIONS

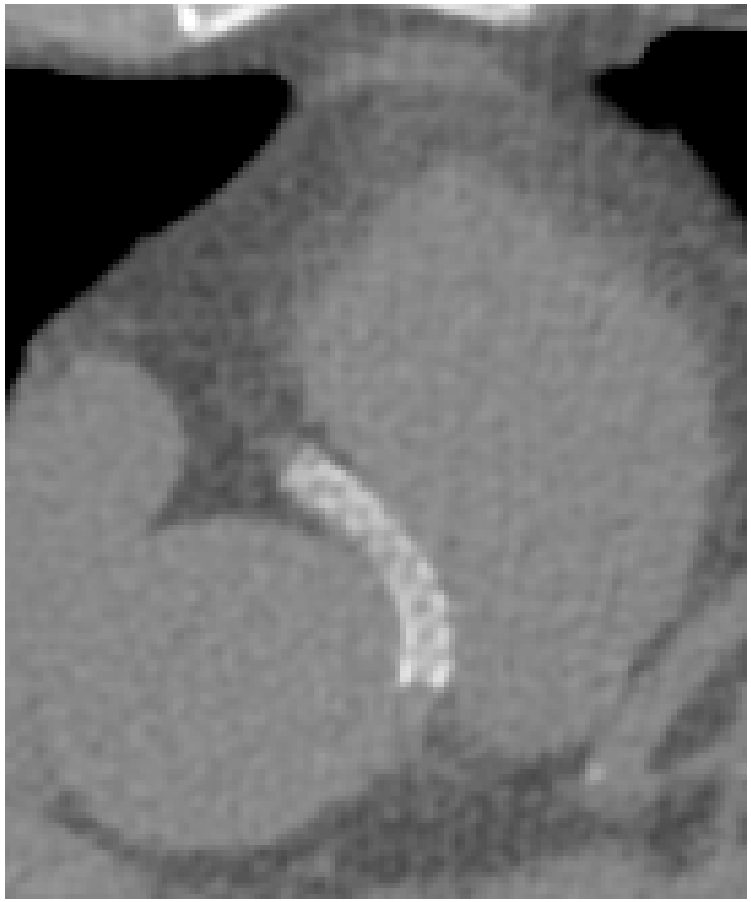
GAP between RECOMMANDATIONS and PRACTICES

- Guidelines focused on young people.
- Recommandations built regardless of age.
- No randomized controlled studies.
- Lack of long-term data after surgical correction.
- Possible failure (stenosis/aneurysm/thrombosis) after surgery.
- Population with very low risk of sudden death (>35 year-old).
- Population with ischemic symptoms (>35 year-old).

CORRECTION INTERVENTIONNELLE

ANGIOPLASTIE





staff ANOCOR

groupe de travail multidisciplinaire de travail sur les ANOmalies congénitales des artères CORonaires

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Phalla Ou (Paris)

ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS

GROUPE HOSPITALIER BICHAT-CLAUDE BERNARD

Algorithme décisionnel (connexions coronaires anormales)

Diagnostic CCA par imagerie invasive ou non invasive

CCA circonflexe

CCA tronc commun ou IVA

CCA coronaire droite

Scanner coronaire si non réalisé

Scanner coronaire si non réalisé

CCA non à risque

CCA à risque

CCA à risque

Pas de traitement spécifique

Test fonctionnel

Imagerie endocoronaire

Test fonctionnel

Imagerie endocoronaire

Eléments décisionnels

- Age / ≤ 30 ans, > 30 ans
- Mode de découverte / Fortuite ?
- Arrêt cardiaque récupéré / Symptomatologie / Lien avec CCA ?
- Ischémie myocardique documentée / Lien avec CCA ?
- Critères anatomiques de gravité
- Maladie athéromateuse associée
- Activité sportive / Type ? / Compétition ?
- Avis éventuel du patient

Décisions proposées

Pas de traitement spécifique

Correction CCA

Traitement anti-ischémique

DAI

Autorisation / Restriction sportive

Angioplastie ou Chirurgie

MERCI

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

8.6. Recommendations for Anomalous Left Coronary Artery From the Pulmonary Artery

Class I

1. In patients with an anomalous left coronary artery from the pulmonary artery (ALCAPA), reconstruction of a dual coronary artery supply should be performed. The surgery should be performed by surgeons with training and expertise in CHD at centers with expertise in the management of anomalous coronary artery origins. (*Level of Evidence: C*)

Warnes CA et al. J Am Coll Cardiol 2008

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary

4.4.5.3. Anomalous Coronary Artery Arising From the PA

Recommendations for Anomalous Coronary Artery Arising From the PA

Referenced studies that support recommendations are summarized in Online Data Supplement 51.

COR	LOE	Recommendations
Therapeutic		
I	B-NR	1. Surgery is recommended for anomalous left coronary artery from the PA.[S4.4.5.3-1–S4.4.5.3-7]
I	C-EO	2. In a symptomatic adult with anomalous right coronary artery from the PA with symptoms attributed to the anomalous coronary, surgery is recommended.
IIa	C-EO	3. Surgery for anomalous right coronary artery from the PA is reasonable in an asymptomatic adult with ventricular dysfunction or with myocardial ischemia attributed to anomalous right coronary artery from the PA.

Stout KK et al. Circulation 2019