Cardiopathies congénitales de l’adulte

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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)
Endorsed by the Association for European Paediatric Cardiology (AEPC)

Baumgartner et al. Eur Heart J 2010

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease
A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

Stout et al. JACC 2018

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease

Khairy et al. Heart Rhythm 2014
Treatment of heart failure in adult congenital heart disease: a position paper of the Working Group of Grown-Up Congenital Heart Disease and the Heart Failure Association of the European Society of Cardiology

Budts et al. Eur Heart J 2016

Chronic Heart Failure in Congenital Heart Disease
A Scientific Statement From the American Heart Association

Stout et al. Circulation 2016

2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC)

Regitz-Zagrosek et al. Eur Heart J 2018
• Notions générales

• Cardiopathies
Épidémiologie (1)

Moons et al. Circulation 2010

Khairy et al. JACC 2010
Figures 2 and 3. The lifetime prevalence of congenital heart disease in children and adults in Quebec, Canada, in 2010. 95% CI indicates the 95% credible interval.

Marelli et al. Circulation 2014
Méconnaissance de la cardiopathie

91 patients, âge médian 17 ans

<table>
<thead>
<tr>
<th>Question</th>
<th>Correct</th>
<th>Incorrect</th>
<th>Does Not Know</th>
<th>Incomplete</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. What is the name of your heart defect?</td>
<td>41 (45%)</td>
<td>9 (10%)</td>
<td>33 (36%)</td>
<td>8 (9%)</td>
</tr>
<tr>
<td>2. Describe or indicate on the diagram where your heart is located.</td>
<td>25 (27%)</td>
<td>12 (13%)</td>
<td>46 (51%)</td>
<td>8 (9%)</td>
</tr>
<tr>
<td>3. How often do you have to come to the clinic for follow-up for your congenital heart disease?</td>
<td>71 (78%)</td>
<td>14 (15%)</td>
<td>6 (7%)</td>
<td>—</td>
</tr>
<tr>
<td>4. What is the main purpose of the follow-up?</td>
<td>42 (46%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>49 (54%)</td>
</tr>
<tr>
<td>5. How has your heart condition been treated to date?</td>
<td>77 (85%)</td>
<td>2 (2%)</td>
<td>4 (4%)</td>
<td>8 (9%)</td>
</tr>
<tr>
<td>6. If you are receiving drug treatment, give the name, dose, schedule, reason or function, most important side effects, and interactions with other drugs or foods.*</td>
<td>8 (53%)</td>
<td>2 (13%)</td>
<td>1 (7%)</td>
<td>4 (27%)</td>
</tr>
<tr>
<td>7. If you experience side effects from your drugs, does this mean you should stop taking them?</td>
<td>37 (41%)</td>
<td>3 (3%)</td>
<td>50 (56%)</td>
<td>—</td>
</tr>
<tr>
<td>8. Do you have to follow a diet? If you answer yes, please indicate the type of diet.</td>
<td>86 (94%)</td>
<td>0 (0%)</td>
<td>5 (6%)</td>
<td>—</td>
</tr>
<tr>
<td>9. Mark all symptoms that may occur if your heart condition deteriorates and for which you have to contact your cardiologist.</td>
<td>8 (9%)</td>
<td>0 (0%)</td>
<td>39 (43%)</td>
<td>44 (48%)</td>
</tr>
<tr>
<td>10. If the congenital cardiologist informs you that everything is all right, does that mean that you do not need further follow-up?</td>
<td>79 (87%)</td>
<td>8 (9%)</td>
<td>4 (4%)</td>
<td>—</td>
</tr>
<tr>
<td>Question</td>
<td>Current</td>
<td>Unknown</td>
<td>Does Not Know</td>
<td></td>
</tr>
<tr>
<td>--------------------------------------------------------------------------</td>
<td>---------</td>
<td>---------</td>
<td>---------------</td>
<td></td>
</tr>
<tr>
<td>11. What is endocarditis?</td>
<td>16 (25%)</td>
<td>9 (15%)</td>
<td>41 (60%)</td>
<td></td>
</tr>
<tr>
<td>12. What is the most typical sign or symptom of endocarditis?</td>
<td>15 (24%)</td>
<td>11 (18%)</td>
<td>43 (68%)</td>
<td></td>
</tr>
<tr>
<td>13. Can you only get endocarditis once in your lifetime?</td>
<td>16 (25%)</td>
<td>14 (23%)</td>
<td>50 (72%)</td>
<td></td>
</tr>
<tr>
<td>14. Do the following factors contribute to the cause of endocarditis?</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Smoking</td>
<td>17 (27%)</td>
<td>18 (30%)</td>
<td>37 (58%)</td>
<td></td>
</tr>
<tr>
<td>Bacterial from dental infections</td>
<td>14 (22%)</td>
<td>12 (20%)</td>
<td>38 (62%)</td>
<td></td>
</tr>
<tr>
<td>Dental discomfort</td>
<td>15 (24%)</td>
<td>15 (25%)</td>
<td>38 (61%)</td>
<td></td>
</tr>
<tr>
<td>Sexual activity</td>
<td>16 (25%)</td>
<td>20 (33%)</td>
<td>40 (62%)</td>
<td></td>
</tr>
<tr>
<td>Wear and use skin care</td>
<td>15 (24%)</td>
<td>17 (28%)</td>
<td>37 (58%)</td>
<td></td>
</tr>
<tr>
<td>Body piercing and tattooing</td>
<td>14 (22%)</td>
<td>11 (18%)</td>
<td>43 (68%)</td>
<td></td>
</tr>
<tr>
<td>15. Because you have a congenital heart disease, should you avoid activity demanding, as you should be careful not to overexert yourself?</td>
<td>21 (32%)</td>
<td>23 (38%)</td>
<td>43 (68%)</td>
<td></td>
</tr>
<tr>
<td>16. Should you have a dental check up at least once a year?</td>
<td>16 (25%)</td>
<td>12 (20%)</td>
<td>50 (72%)</td>
<td></td>
</tr>
<tr>
<td>17. Should you make antibiotics before every visit to the Journal?</td>
<td>16 (25%)</td>
<td>12 (20%)</td>
<td>50 (72%)</td>
<td></td>
</tr>
<tr>
<td>18. The following genes need visual assessment?</td>
<td>16 (25%)</td>
<td>15 (24%)</td>
<td>45 (71%)</td>
<td></td>
</tr>
<tr>
<td>19. Should you have a visit at least once a year?</td>
<td>17 (27%)</td>
<td>14 (23%)</td>
<td>43 (68%)</td>
<td></td>
</tr>
<tr>
<td>20. Is teaching more beneficial for patients with congenital heart disease than for other people?</td>
<td>21 (32%)</td>
<td>19 (31%)</td>
<td>42 (66%)</td>
<td></td>
</tr>
<tr>
<td>21. Is continuing alcohol a little more beneficial for patients with congenital heart disease than for other people?</td>
<td>23 (36%)</td>
<td>21 (34%)</td>
<td>36 (58%)</td>
<td></td>
</tr>
<tr>
<td>22. Can you take part in competitive sports requiring daily training?</td>
<td>35 (59%)</td>
<td>42 (66%)</td>
<td>14 (21%)</td>
<td></td>
</tr>
<tr>
<td>23. Should you choose an occupation that is not too physically demanding, as you should be careful not to overexert yourself?</td>
<td>68 (75%)</td>
<td>11 (12%)</td>
<td>12 (13%)</td>
<td></td>
</tr>
<tr>
<td>24. In terms of sexual physical effort, can you do all that you feel you are able to do?</td>
<td>71 (78%)</td>
<td>1 (1%)</td>
<td>19 (21%)</td>
<td></td>
</tr>
<tr>
<td>25. What is the chance that your children will have congenital heart disease?</td>
<td>18 (20%)</td>
<td>24 (26%)</td>
<td>49 (54%)</td>
<td></td>
</tr>
<tr>
<td>26. Which contraceptives are the most advisable for you to use in light of your congenital heart disease? (only for women)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Contraceptive pill</td>
<td>15 (35%)</td>
<td>—</td>
<td>28 (65%)</td>
<td></td>
</tr>
<tr>
<td>Intrauterine device</td>
<td>6 (14%)</td>
<td>3 (7%)</td>
<td>34 (79%)</td>
<td></td>
</tr>
<tr>
<td>27. Are you at risk of deterioration during pregnancy? (only for women)</td>
<td>5 (12%)</td>
<td>11 (25%)</td>
<td>27 (63%)</td>
<td></td>
</tr>
</tbody>
</table>

Van Deyk et al. Am J Cardiol 2010
643 patients, nés en 1983, vivants à 22 ans et avec diagnostic de CHD avant l’âge de 6 ans, au Canada
13% de cardiopathies complexes, 61% de simples shunts

**Facteurs prédicifs:** sexe masculin, lésion peu sévère, suivi dans un centre non universitaire
Perdus de vue (2)

21% des patients avec cardiopathie complexe perdus de vue après 18 ans
The Adult With Congenital Heart Disease
Born to Be Bad?
Carole A. Warnes, MD, MRCP, FACC

The perception of “cure” is fostered by the surgical description “total correction,” which is applied to many operative repairs of complex congenital anomalies. In reality, there is almost no surgical cure for congenital heart disease, perhaps with the exception of a successfully ligated and divided ductus arteriosus. All other repaired lesions have the potential for residua and sequelae.
Quelles conséquences? (2)

Hospitalisations en urgence

Gurvitz et al. JACC 2007
Quelles conséquences? (3)

Complications évitables

Coarctation aortique

41% HTA

De Bono et al. Int J Cardiol 2012

Fermeture spontanée

8 (4 %)

Mortalité

2 (1 %)

Insuffisance valvulaire aortique

53 (26 %)

Arythmies V

1 (0.5 %)

Troubles de conduction

2 (1 %)

Endocardite infectieuse

8 (4 %)

Oclusion instrumentale

15 (7 %)

CIV pm

Tétralogie de Fallot

Soufflet et al. Am J Cardiol 2010

Pour normaliser les volumes VD en post-opératoire: RVP quand VTDVD <160 ml/m² ou VTSVD<82 ml/m²

Oosterhof et al. Circulation 2007
Les congénitaux adultes sont-ils seulement de grands enfants?

- Changement de problèmes médicaux
- Cardiopathies acquises surajoutées (ischémie coronaire, HTA...)
- Techniques d’examen différentes
- Considérations sociales: emploi, assurance, revenus, logements, prêts...
- Contraception et grossesse
Transition-Principes

- Support institutionnel
- Coordinateur de transition
- Plan de transition écrit
- Faciliter la communication directe patient-soignant
- Education permanente des parents et du patient
- Consultations spécifiques de transition
Recommandations européennes
Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association

For Adolescents

1. The timing of transition should be guided by emotional maturity and developmental level (as opposed to chronological age) for transition planning (Class I; Level of Evidence C).
2. The adolescent should be engaged in transition planning (Class I; Level of Evidence C).
3. The adolescent should be asked about their understanding of their disease in relation to their current health status, restrictions on activities, and future goals (Class I; Level of Evidence C).
4. The adolescent should be encouraged to share concerns about QOL issues (physical restrictions, school, peers, social relationships) (Class I; Level of Evidence C).
5. The adolescent’s fears and concerns should be acknowledged in an empathetic, nonjudgmental manner (Class I; Level of Evidence C).
6. The pediatric cardiology provider should initiate and work together with the adolescent on a transition plan using a transition resource binder and/or health “passport” (Class I; Level of Evidence C).
7. Providers should begin to direct health discussions more toward the adolescent than the parent (Class I; Level of Evidence C).

For Parents

1. The pediatric cardiology provider should initiate discussions on transition planning and partner with parents in the process (Class I; Level of Evidence C).
2. The pediatric cardiology provider should solicit information about parental perceptions of their child’s QOL (Class I; Level of Evidence C).
Quelles thématiques aborder ?

• Connaissance de la cardiopathie
• Modalités de suivi et enjeux
• Conseil génétique, risque de récidive
• Sexualité, contraception, grossesse
• Activité physique possible, limites
• Apports alimentaires
• Tabac, alcool, drogues
• Emplois possibles
• Assurances, prêts
• Support psychologique?
• Notions générales
• Cardiopathies
Mme Marine A, ddn 18/07/1981

• Tétralogie de Fallot

• Cure complète le 11 septembre 1984 (large patch d’élargissement infundibulo-pulmonaire, fermeture de CIV)

• Suivi aléatoire (asymptomatique)

• 1 grossesse sans complications en 2008

• Revient avant nouvelle grossesse...

• Se dit asymptomatique

• BDC réguliers, SS 2/6 RSG, SD court 3/6 RSG, absence d’IVD
Que lui dites-vous?

• Aucun problème!
• Nécessité d’un bilan préalable avec ETT et HolterECG
• Bilan plus complet
• Contre-indication absolue
• Cathétérisme cardiaque
Revalvulation pulmonaire chirurgicale (homogreffe) et annuloplastie tricuspidie
- 1 an après : grossesse

Brickner et al. *NEJM* 2000