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Adherence to guidelines in the clinical care for adults with congenital heart disease: The Euro Heart Survey on Adult Congenital Heart Disease

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Aims To investigate the role of guidelines in structuring the clinical care for adult patients with congenital heart disease (CHD), and to assess adherence to the guidelines in Europe.
Methods and results A selected number of current guidelines were chosen pertaining to operative procedures, investigations, and the use of medication ('interventions'). The source for this analysis was the database of the Euro Heart Survey on adult CHD, which contains retrospectively collected data on 4110 patients followed-up for a median of 5.1 years. For each guideline investigated, patients were selected from the database for whom the particular guideline was relevant. The selected cases were classified according to two criteria: was there an indication for the particular intervention and did the intervention take place? In this manner, cases of 'undue treatment' and 'insufficient treatment' were identified. Adherence to guidelines was found to be good in the case of operative procedures and prophylactic drug treatment. However, regarding diagnostic procedures there had been adherence to guidelines in only slightly more than half of the cases.
Conclusion Guidelines have an important role in the actual clinical care of adults with CHD. However, large outcome studies are needed to develop more precise guidelines.

Introduction

One of the central themes of the Euro Heart surveys is the adherence to guidelines in day-to-day clinical decision-making. To assess the role in clinical practice of recommendations based on expert consensus was an important aim of the recently completed Euro Heart Survey on congenital heart disease (CHD) in adults. In the relatively new field of adult CHD—in contrast to most other fields of cardiology—there have hardly been any large randomized trials to validate principles of management and compare treatments. In such circumstances, guidelines may yet fulfil an important role in formulating what constitutes optimal treatment, in summarizing clinical consensus, and in structuring clinical practice and research. In adult CHD, guidelines are especially important for decision-making and proper timing of (re-)intervention, for determining the optimal diagnostic modality and frequency for patient follow-up, and for the prescription of (prophylactic) medication. If it is found that guidelines are not adhered to in actual practice, it means that either education of practitioners needs to be improved, or that the guidelines themselves are deficient and need to be adapted.

Two major sets of guidelines have recently been published in the field: the Canadian Cardiovascular Society/American College of Cardiology guidelines[1–4] and the guidelines of the European Society of Cardiology.[5] In this report, we describe the adherence to guidelines as it appeared from the data that were collected as part of the European adults CHD survey.

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Methods

The general methods followed in collecting the data for this survey have been described previously.[6] Briefly, the patient population consisted of patients older than 17 years of age who had one of the following congenital cardiac conditions (referred to as 'defects' in the following): atrial septal defect II (ASD), ventricular septal defect (VSD), tetralogy of Fallot (ToF), complete transposition of the great arteries (TGA), aortic coarctation (CoA), Marfan syndrome, Fontan circulation, or cyanotic disease. Consecutive patients fulfilling the inclusion criteria (see below) were included retrospectively at their first visit to the outpatient clinic in 1998 (or else 1999 or 2000), and their follow-up data until December 31, 2003, were recorded reviewing the medical chart. The database was opened for data entry on July 1, 2003, and closed on April 30, 2004. Data were derived from the patient files by trained nurses or physicians and then entered into an electronic case record file (CRF). Data collected included clinical characteristics, diagnostics and interventions performed. Any significant changes in the clinical condition that occurred during follow-up were recorded. Separate sections were devoted to interventional procedures, requiring both details regarding previous interventions, and those performed during the follow-up period.

Adherence to guidelines was evaluated for three separate categories of guidelines: guidelines for diagnostic procedures, operative- and catheter-based interventions, and drug treatment. Guidelines to be chosen for analysis was determined by the following criteria: relevance to clinical decision-making; relative simplicity; and fitness of the required data for CRF-based evaluation. An overview of the evaluated guidelines is shown in *Tables 1–3*. In order to assess adherence to these guidelines, the patients for whom the guidelines were relevant were selected from the database. Next, the cases of these patients were entered into an analysis that we shall describe separately for the different categories of guidelines.

Table 1 Overview of evaluated guidelines on diagnostic investigations

Coronary angiography prior to surgery
Marfan patients >50 years of age
Other patients >40 years of age
Imaging of the thoracic aorta in repaired aortic coarctation
Diagnostic work-up should include imaging of the aorta by MRI, DSA, or angiography
Exercise testing in aortic coarctation
Diagnostic work-up should include exercise testing
Imaging of the abdominal aorta in Marfan patients
During follow-up, the entire aorta should be regularly evaluated with echocardiography, MRI, CT, and/or abdominal echo

Diagnostic investigations

Coronary angiography prior to surgery

All patients who underwent surgery were selected from the database and classified according to two criteria: whether or not

Table 2 Overview of evaluated guidelines on interventions

ASD closure
A 'significant' ASD warrants intervention
A 'significant' ASD:
causes volume or pressure overload (i.e. moderate/severe RV overload, PAP >35 mmHg, or LR shunt >1.5:1)
may cause exercise limitation (NYHA class >I)
may cause atrial arrhythmias
may cause paradoxical embolism
VSD closure
The following conditions may warrant VSD closure:
The presence of a 'significant' VSD (i.e. moderate/severe LV overload, or PAP >50 mmHg, or LR shunt >2:1, or NYHA class >I)
The presence of a 'significant' RVOT obstruction (i.e. gradient >50 mmHg, or judged to be moderate/severe)
The presence of a perimembranous VSD with moderate/severe aortic regurgitation
The presence of a subaortic VSD in combination with aortic valve prolapse
A history of endocarditis
Pulmonary valve replacement in Fallot
The presence of severe pulmonary regurgitation may warrant pulmonary valve replacement if it goes together with one of the following:
Progressive RV enlargement
Progressive tricuspid valvular regurgitation
NYHA class > I due to the defect
Arrhythmias
(Re-)coarctectomy in aortic coarctation
Presence of a 'significant' (re-) coarctation may warrant intervention. A 'significant' re-coarctation gives rise to hypertension (140/90), plus one of the following:
'pull-back' gradient >20 mmHg; an arm-leg gradient >30 mmHg
Aortic root replacement in Marfan patients
The following situations warrant aortic root replacement:
Aortic root diameter >55 mmHg
Aortic root diameter >50 mmHg, plus:
Family history dissections, or
Growth rate >2 mm/year, or
Severe mitral or aortic valvular regurgitation
Aortic root diameter >45 mm in case valve-sparing surgery is planned
Aortic root dissection

coronary angiography was performed, and whether or not they met the (age) criteria specified in the guideline. This led to four possibilities, two of which were classified as undue-, respectively, insufficient use of diagnostic investigations. An example of this decision process is shown in *Figure 1*.

Imaging of the aorta and exercise testing

As the guidelines do not specify how frequently these screening procedures should be performed, it was determined in which percentages of CoA, respectively Marfan patients, these procedures had been performed after 3, 4, and 5 years of follow-up. Imaging was defined as follows: in CoA, an investigation by either magnetic resonance imaging, or digital subtraction angiography; in Marfan syndrome, an investigation by either magnetic resonance imaging, or computed tomography scan, or abdominal echography. Numbers of patients still under follow-up after 3, 4, and 5 years are given as counts; percentages are followed by 95% confidence intervals (CI).

Operative- and catheter-based interventions

Essentially, the guidelines for operative- and catheter-based interventions specify that a particular intervention should be performed when one or more conditions in the status of the patient are fulfilled. In the following, we refer to this situation as the existence of an indication.

We first determined to which patients the particular guideline applied. These were, respectively, patients with an open ASD at study entry; patients with an open VSD at study entry; all Fallot patients; all CoA patients; Marfan patients who had not undergone an intervention before inclusion. Next, for each of these patients it was decided whether all data items were available that were necessary to evaluate whether or not an indication existed and whether or not an intervention had occurred. Patients for whom these data were lacking or insufficient were excluded from further analysis. The remaining cases were subjected to the following assessment.

It was determined if at one point of time during follow-up an indication existed, and whether or not the patient had undergone the particular intervention. If the answers to these two questions were concordant, the patient was considered to have been treated according to the guidelines. Discordant answers could result from either one of two possibilities that were further analysed as follows:

(i) The patients with an indication who did not undergo an intervention. The files of these patients were screened by the investigators (BM, PE, EB, JT: the 'panel') for data that might explain why an intervention was not mentioned in their record. This was done first by running through a list of reasons that were explicitly collected on the CRF. This included the following possibilities: 'scheduled for intervention', 'referred' (the design of the survey and anonymity requirements did not allow tracing the fate of such patients), 'patient refusal', 'lost to follow-up', 'deceased', 'high-risk', 'mentally retarded', or 'symptoms owing to other pathology'. The remaining cases were further evaluated according to whether one of the following situations applied: (a) the indication criteria were met only marginally or there were contra-indications not mentioned in the current guideline. This could lead to the consideration that non-intervention was justified after all; (b) essential data were lacking after all (which had only become apparent after a closer scrutiny of the case record). The cases left over after this 'sieving' process were considered as having not been treated according to the guidelines ('insufficient treatment'). After this analysis, each case was classified as 'explained' (scheduled for intervention, referred, patient refusal, symptoms due to other pathology, high-risk, mentally retarded, or otherwise justified by panel), or 'unexplained'; the unexplained cases included those who had deceased or were lost to follow-up (we considered that, in principle, we could not know whether or not these patients had been treated properly, as their death or being lost to follow-up could either have prevented intervention or have been the result of non-intervention), patients for whom data were found to be lacking after all, and, finally, the patients who, according to this analysis, had not been treated in accordance with the guidelines.

(ii) The patients who underwent a relevant intervention, but without an indication. Similar to the above, a reason was sought by the investigators that could explain the intervention. Acceptable reasons could be that the patient suffered from the more elusive symptoms mentioned in the guidelines as warranting, but not necessarily requiring an intervention in all cases; or

Table 3 Overview of evaluated guidelines on medication

β-blocking agents in Marfan
All patients, unless intolerable side effects
Anti-coagulants for mechanical valves
All patients with a mechanical valve should be on anti-coagulants
Anti-thrombotics for arrhythmias in Fontan
Fontan patients with atrial arrhythmias should be anti-coagulated long-term

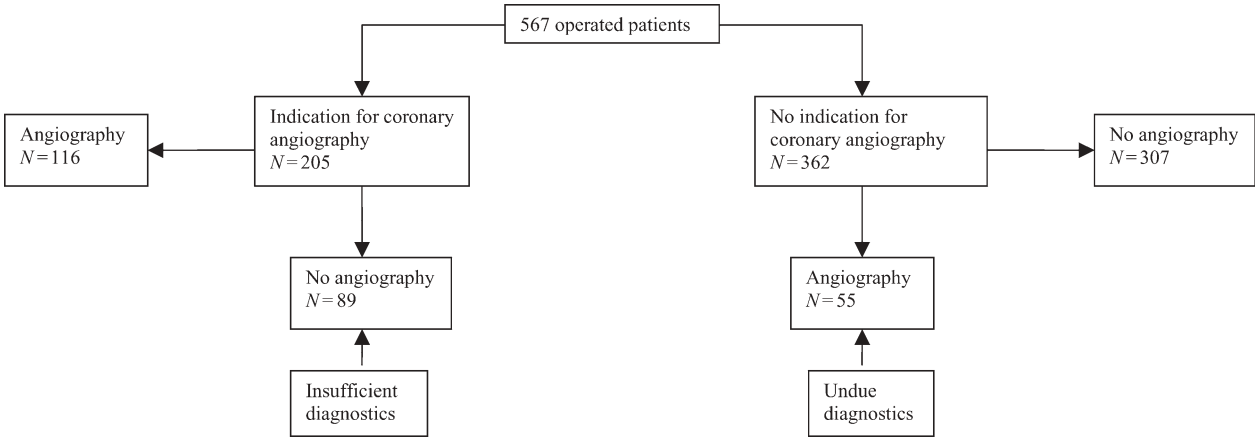

Figure 1 Flow chart showing which numbers of operated patients with, respectively without, an (age-based) indication for pre-operative coronary angiography actually underwent coronary angiography.

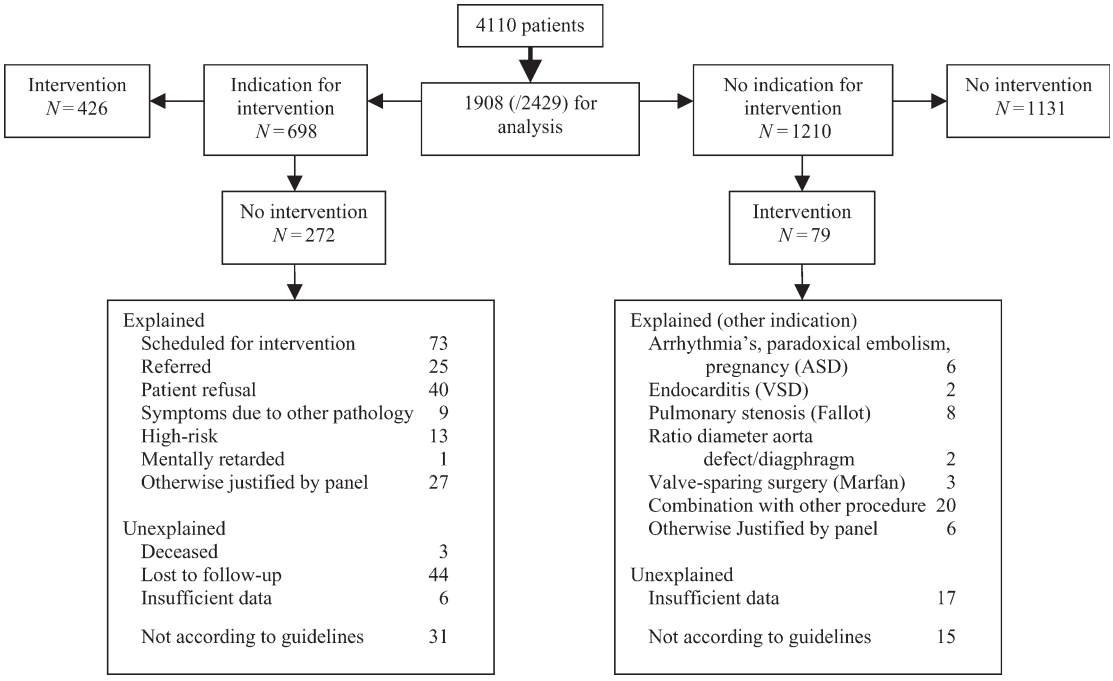

Figure 2 Flow chart showing both the method used and the results obtained in assessing adherence to guidelines for surgical or catheter-based interventions: ASD closure, VSD closure, pulmonary valve replacement in ToF, (re-)coarctectomy in CoA, and aortic root replacement in Marfan syndrome).

the patient could have an alternative indication, not mentioned in the specific guideline; or the intervention was combined with another procedure. This, again, resulted in a classification of each of these cases as either 'explained' (meaning that intervention was justified after all) or 'unexplained'. Some of the unexplained cases were judged to be unexplained as essential data was lacking after closer scrutiny of the record. The remaining cases were considered as treatment not according to the guideline ('undue treatment').

The process of evaluating adherence to guidelines for interventions may be visualized by referring to *Figure 2*, which summarizes the results of the analyses (see Results section below).

Drug treatment

The CRF contained a section devoted to drug use at inclusion and during follow-up. When patients were started on medication during follow-up, entry of starting dates was requested. The Kaplan–Meier method was used to estimate the percentages (95% CI) of patients who were using medication at the end of the follow-up period.

All statistical calculations were performed using the statistical package SPSS, version 12.01.

Results

The database of the Euro Heart Survey on adult CHD contains data of 4110 patients. Numbers of patients per defect and baseline characteristics of the patients are displayed in *Table 4*. As explained earlier (Methods section), from among these patients all patients were selected to whom one of the assessed guidelines applied. *Table 5* provides an overview of the numbers of patients selected for each guideline as well as the proportion of those for whom sufficient data for evaluation were available.

Table 4 Baseline characteristics of all patients

	Inclusion (*n*)	Age[a]	Females (%)
Atrial septal defect II	882	39 (25–52)	67
Ventricular septal defect	628	27 (20–36)	53
Tetralogy of Fallot	811	26 (21–34)	48
Aortic coarctation	551	26 (21–35)	39
Transposition great arteries	363	23 (20–27)	39
Marfan syndrome	287	29 (22–38)	50
Fontan circulation	198	23 (19–28)	45
Cyanotic defect	390	29 (23–37)	62
Overall	4110	27 (23–37)	52

[a]Figures represent medians (inter-quartile range).

Diagnostic investigations

Coronary angiography prior to surgery

In total, there were 608 patients who underwent an operation. However, we could not include 41 patients who underwent coronary angiography but who reached their 40th birthday (50th in the case of Marfan patients) during follow-up, as it was not known whether the angiography was performed before or after that date. Thus, 567 patients remained for the analysis, the results of which are displayed in *Figure 1*. On the basis of these findings, the group of operated patients with an indication for coronary angiography but who did not undergo angiography was compared with the group with an indication who did undergo angiography. Disregarding the Marfan patients, the patients who did not undergo angiography were significantly younger than patients who did [median 46 years (inter-quartile range: 43–57) vs. 53 (47–62) years; $P < 0.001$, as assessed by the Mann–Whitney test].

Table 5 Patients included per guideline assessed

Diagnostics	*n*/*m*[a]	Interventions	*n*/*m*	Drug treatment	*n*/*m*
Imaging thoracic aorta in CoA	551/551	Closure of ASD	495/504	β-blockers in Marfan	287/287
Exercise testing in CoA	551/551	Closure of VSD	247/352	Anti-coagulants for mechanical valves	316/316
Imaging abdominal aorta in Marfan	286/287	PVR in Tetralogy of Fallot	547/811	Anti-thrombotics for arrhythmias in Fontan	117/117
Coronary angiography prior to surgery	567/608	(Re-)coarctectomy in CoA	411/551		
		ARR in the Marfan syndrome	208/211		

PVR, pulmonary valve replacement; ARR, aortic root replacement.
[a]The figures represent the total number of patients to which the guideline applied (*m*) and the number of patients with sufficient data for analysis (*n*).

Imaging of thoracic aorta/exercise testing in CoA
The results for the guidelines on imaging of the thoracic aorta and exercise testing in the patients with CoA, as well as the numbers of patients with at least 3, 4, and 5 years of follow-up respectively, are given in *Table 6*.

Imaging of the abdominal aorta in Marfan
Likewise, provided in *Table 6* are the proportions of patients who had undergone imaging of the abdominal aorta at 3, 4, and 5 years respectively, as well as the number of patients still under follow-up at these times.

Surgical- and catheter-based interventions

The aggregate results of the analyses as described in the Methods section is shown in *Figure 2*. For each guideline separately, *Table 7* gives the number of patients with an indication, followed by the number of cases of 'undue treatment', respectively 'insufficient treatment'. Overall, 31 of the 698 patients with an indication were considered to have been treated insufficiently, i.e. 4% (95% CI 3–6%); and 15 of the 1210 patients without an indication had an unwarranted intervention, i.e. 1% (1–2%).

Please note that in the Discussion section some details on individual cases are mentioned. These serve the purpose of illustrating some issues in the interpretation and functioning of guidelines, and are not spelled out here.

Drug treatment

β-blocking agents in Marfan
Results regarding β-blocking agents in Marfan syndrome, anti-coagulants for mechanical valves (136 patients with a mechanical valve were identified) and anti-thrombotic therapy for arrhythmias in Fontan are given in *Table 8*.

Discussion

Even though the care for adult CHD patients is a young discipline, guidelines have an important role in structuring clinical practice. The results of the analyses presented here confirm that the actual adherence to the proposed standards in clinical care is good with regard to the surgical- and catheter-based interventions and the prescription of prophylactic drugs, but not quite up to targets in the case of diagnostic investigations. As far as surgical- and catheter-based interventions are concerned, adherence to guidelines is particularly good to relatively simple and well-researched clinical guidelines, namely aortic root replacement in the Marfan syndrome, and ASD and VSD closure.

The method that was used to assess adherence to guidelines was to 'filter out' the patients who were not treated strictly according to the guidelines, and to subject these cases to a further scrutiny of the patient file for additional clinical information that could explain apparent deviation from the guideline. The logic behind this last step in the sieving process was the assumption that there is a 'grey area' in which the guideline is not decisive. In this discussion, particular attention will be devoted to these borderline cases, as they are the most instructive in evaluating the appropriateness of the current guidelines.

Diagnostic investigations

Most adult CHD patients are under regular follow-up. Screening and monitoring therefore are key elements in optimizing care.

Pre-operative coronary angiography is indicated in older patients to exclude coronary artery disease that might complicate the peri-operative course. We found that almost half of the patients above 40 years of age (50 in the case of the Marfan syndrome, because of the risk of compromising the fragile aortic root) did not undergo coronary angiography before surgery. The fact that the mean age was significantly lower in the patients who did not undergo angiography seems to indicate that in practice a higher age limit is observed. It should be noted that the prevalence of coronary artery disease as diagnosed by coronary angiography is below 4% for asymptomatic patients aged 40–70 years[7] and does not seem to increase sharply between 40 and 50 years of age. The evidential basis for a lower limit of an age of 40 as criterion for performing coronary angiography is rather weak. Therefore, this guideline might be a candidate for revision.

The other diagnostic procedures assessed pose a problem of a somewhat different nature. All pertain to screening and monitoring, i.e. serial measurements. Yet, the guidelines are silent as to how frequently investigations should be done. We have therefore considered periods of follow-up of 3, 4, and 5 years, assuming that the aortic diameters should be determined at least once in every 5 years. The fact that we found hardly any differences between follow-up of 3 and 5 years suggests that either aortic diameters are being monitored at regular time-intervals of at most a few years, or that they are monitored infrequently, or not at all. In any case, the outcome of our analysis should be

Table 6 Diagnostic investigations

	Aortic coarctation				Marfan	
	X-ECG ($n = 551$)		Imaging thoracic aorta ($n = 551$)		Imaging abdominal aorta ($n = 286$)	
3 years follow-up	$n = 478$	52% (47–56)	$n = 478$	54% (50–58)	$n = 261$	66% (60–71)
4 years follow-up	$n = 405$	53% (48–58)	$n = 405$	55% (50–59)	$n = 223$	70% (64–76)
5 years follow-up	$n = 325$	52% (47–57)	$n = 325$	55% (49–60)	$n = 182$	73% (66–79)

Figures represent percentages (95% confidence interval) preceded by the number of patients with 3, 4, and 5 years of follow-up, respectively.

Table 7 Adherence to the guidelines for surgical/catheter-based interventions

	No. of patients with indication	Cases of undue treatment	Cases of insufficient treatment
Closure of ASD	434 /495	1	3
Closure of VSD	80 /247	1	1
PVR in Tetralogy of Fallot	65 /547	5	10
(Re-)coarctectomy in CoA	86 /411	6	14
ARR in the Marfan syndrome	33 /208	2	3
Overall	698 /1902	15 (1%)[a]	31 (4%)[b]

PVR, pulmonary valve replacement: ARR, aortic root replacement.

Figures represent counts. The denominators in the first column represent the number of patients to whom the guideline applies and for whom sufficient data was available.

[a]Expressed as percentage of patients without an indication (95% confidence interval: 1–2%).

[b]Expressed as percentage of patients with an indication (95% confidence interval 3–6%).

Table 8 Prophylactic medication

	Using mediation[a] (%)
β-blockers in Marfan	76 (71–82)
Anti-coagulants for mechanical valves	95 (91–99)
Anti-thrombotics for arrhythmias in Fontan	91 (83–99)

[a]Figures refer to the percentages of patients (95% confidence interval) using medication at baseline or started at some time during follow-up, as estimated with the Kaplan–Meier method.

the cause for concern that this guideline is insufficiently put into practice. As availability of imaging equipment is still limited, no distinction was made regarding the mode of imaging. However, it is to be expected that future research results will favour particular forms of imaging.

Surgical- and catheter-based interventions

In contrast to most other congenital heart defects, ASDs often manifest themselves for the first time in adulthood. In most specialized centres, the evaluation of open ASDs in adults is an important part of their care. The relatively long clinical experience with the management of ASDs has led to guidelines that are quite clear and well supported. Surgical repair of ASDs in childhood is known to be associated with excellent outcomes.[8] Because now almost all ASDs discovered in childhood are closed, we may assume that in most cases of an open ASD in our database the defect had remained undiscovered during childhood. The risks and benefits of closure later in life have been the subject of debate. In only one prospective randomized study, surgical treatment was compared with medical treatment in adults.[9] Despite the lack of hard evidence, the results of several retrospective follow-up studies[9–12] have led to the almost uniform consensus that a 'significant' ASD when it becomes manifest in adulthood should be closed. Our analysis seems to confirm that the guideline is closely adhered to. However, the precise conditions favouring closure vs. conservative treatment have not been clearly defined. Moreover, closure of ASDs by a catheter-based technique appears to be associated with fewer/less severe complications in older patients, although long-term follow-up is not yet available. Hence, this current trend might necessitate a revision of the guideline.

In our cohort, a shunt with a Qp:Qs ratio $> 1.5:1$ and/or pulmonary hypertension were major indications for treatment. It should be noted, however, that a cut-off of 1.5:1 for the shunt ratio is not always observed in the literature. Thus, Attie *et al.*[9] used a ratio greater than 1.7:1, established by right heart catheterization, as an inclusion criterion for their study.

Apart from these haemodynamic criteria, the guidelines mention a history of arrhythmias and/or paradoxical embolism as possible indications for ASD closure. However, it has been shown that ASD closure is not an effective remedy for arrhythmias.[10,11] In our cohort, there were two patients with atrial arrhythmias as the sole reason for ASD closure. In addition, there were seven patients in whom paradoxical embolism was the sole reason for closure.

Compared with ASDs, only few VSDs needed to be closed [36 of 352 unclosed VSDs (10%), compared with 296 of 504 unclosed ASDs (59%)], which is not surprising given the fact that most VSDs are discovered in childhood. It is known that the long-term outcome is good after VSD closure in

infancy and childhood.[13] On the other hand, an unclosed significant defect may lead to substantial pathology later in life, in particular pulmonary hypertension and left ventricle overload. We found no patients who should certainly have been operated but who were not. There were, however, several cases that give food for thought. In particular, the role of a history of endocarditis is unclear. In three cases, this was the only reason for operating, while on the other hand 12 unoperated patients had a history of endocarditis. There also is the question whether patients with only one of the indications for intervention should always be treated accordingly. For example, two patients had only moderate left ventricular volume overload, and two patients were in NYHA functional class II due to the VSD, but did not qualify for any of the other criteria. On the other side of the spectrum, a patient should be mentioned who was in NYHA class IV and had a left ventricular ejection fraction of 31%, but was not operated. Would this patient have benefited from closure of the VSD, or would an operation have further compromised the left ventricular function and provoked heart failure? In other words: should the guideline specify when the risks of operation outweigh the benefits?

Probably, the least well-established guideline is that for pulmonary valve replacement after correction of the ToF. After corrective repair of Fallot, re-intervention was necessary in about 10% of patients over a 20-year follow-up period.[14] In most cases, the intervention concerned the right ventricular outflow tract. One recent study reported that in a population of 171 patients with a mean follow-up of approximately 10 years, 9% of the patients underwent pulmonary valve repair.[15] Some degree of pulmonary valve insufficiency develops in almost all patients. It is generally agreed that in the more severe forms of pulmonary regurgitation, pulmonary valve replacement is indicated. However, the timing of the intervention is still an important matter of debate and research.[16,17] Therefore, it must be concluded that the indications as set forth in this guideline are subject to alternative interpretation and are likely to be revised as more outcome data become available. Not surprisingly, we found more departures from this guideline than from those in the other cases. Deviations may be categorized as follows. First, in several cases pulmonary stenosis and not pulmonary insufficiency was the probable reason for pulmonary valve replacement. In other cases, moderate (and not severe) pulmonary regurgitation occurred in combination with one of the other criteria. This reflects the fact that it may be difficult to quantify the severity of PI. In one case, moderate PI in combination with a right ventricular outflow tract aneurysm (not mentioned in the guideline) probably was the reason for intervention. But even after classifying all these cases as 'acceptable', 15 cases remained in which treatment was incorrect, according to our appraisal (10 cases of 'insufficient treatment' and five cases of 'undue treatment').

Compared with the guidelines discussed earlier, which pertain to often complex clinical decision-making, the guideline for aortic root replacement in Marfan is rather straightforward and provides clear-cut criteria that can be readily established. The observation that dissection/rupture of the aorta in Marfan syndrome is in many cases preceded by a widening of the aortic root, has led to the strategy of prophylactic replacement of the aortic root. Cut-off values have been proposed and found to be satisfactory.[18] We found only three cases in which aortic root replacement was not performed when it should have been, and two cases in which it was performed while the aortic root diameter was below the threshold. A further indication for the impact of this guideline is the fact that in most cases aortic root diameters were available. Yet, it should be noted that when aortic root regurgitation is present, it may be a reason to replace the aortic root earlier (a few cases in our database). Also, improvements of surgical techniques, in particular valve-sparing techniques, may shift the benefit/risk ratio and provide a rationale for earlier intervention.

The guideline for (re-)coarctectomy likewise relates to monitoring of aortic dimensions. After repair of CoA, residual or re-stenosis at the site of repair often necessitates a repeat intervention,[19] while untreated patients with (mild) CoA may become symptomatic later in life. The main reason to intervene in these cases is to attenuate the deleterious effects of the systemic arterial hypertension that goes with coarctation. These are such that, except in the case of very mild obstructions, interventions are indicated. The crucial haemodynamic feature that is used for diagnostic and monitoring purposes is the difference in blood pressure in the upper and the lower part of the body. This can be determined invasively by catheter. However, invasive diagnostics have been largely replaced by non-invasive imaging modalities, in particular Doppler echocardiography ('sawtooth' phenomenon) and magnetic resonance imaging. Unfortunately, these methods do not directly assess the haemodynamic severity of the stenosis. The requirement of the presence of 'resting or exercise-induced hypertension and a resting arm/leg blood pressure gradient >30 mmHg' is therefore a criterion that is, in a sense, a compromise.

Judging from our data, actual treatment tends to be more conservative than recommended in this guideline. We found 16 cases in which the intervention criteria were fulfilled but in which no intervention took place, vs. five cases in which an unwarranted intervention was performed. In one of these cases the criterion for intervention was the ratio of aortic diameter at the stenosis site over the diameter at the diaphragmatic level (<0.7), which is an accepted criterion. It should further be noted that the untreated cases might include patients who were inoperable for anatomical reasons (hypoplastic arch).

This tendency towards a more conservative approach is especially noteworthy given the fact that recent research findings indicate that treatment should be even more aggressive than as recommended by the current guideline, in particular, as the advent of transcatheter techniques will reduce interventional risks.[20]

Medication

In general, the repertoire of drugs used in adult CHD is not different from that in other fields of cardiology. However, the indications for prescribing medication may be different. One example is the prophylactic prescription of β-blockers to prevent aortic dissection in Marfan patients.[21] The fact that more than 70% of the Marfan patients in the database were using β-blockers shows that this guideline is fairly well established, considering that the existence of side effects and patient reluctance to use β-blockers render the target of 100% unrealistic.

The use of anti-thrombotics after mechanical valve implantation is a general principle in cardiology.[22] Our finding that almost 10% of patients in this category were not using any form of anti-coagulation is therefore slightly alarming.

Prevention of thrombo-embolic events is also necessary in patients with Fontan circulation when they suffer from arrhythmias. As the prevalence of supraventricular arrhythmias is very high, this concerns a large proportion of the Fontan patients. Unfortunately, the Fontan circulation may also disturb the coagulation profile in such a manner that bleeding disorders result.[23] Anti-coagulation management in Fontan patients therefore, remains a major challenge that future guidelines will need to address.

Limitations

Evaluation of the adherence to guidelines in this survey did not have the character of an 'audit' and cannot be considered as an assessment of the 'quality of care'. Participation was on a voluntary basis, and data-entry was done by the participants in this survey, who, in many cases, included their own patients, with no hard guarantee of accuracy and reliability. Moreover, we based our assessment on data that were collected by means of a standardized electronic CRF. This imposed limits on the amount of detail with which we could evaluate each case. In particular, we were not able to include the full process of clinical decision-making with all its subtleties. On the one hand, we were not able to further analyse patients who were lost to follow-up, were referred to another centre, or died. On the other hand, we were not able to give the treating cardiologist a 'fair hearing' in case of apparent non-adherence to a guideline.

Conclusion

This survey shows that guidelines have gained an important role in the care for adult patients with CHD. Actual adherence to the proposed standards was found to be generally good, although in some aspects of clinical care, in particular the performance of diagnostic investigations at regular intervals, there was a lack of compliance, which is a cause for concern. As guidelines mostly incorporate previously established rules of good practice, also in a young discipline such as adult CHD treatment, recommendations should be formulated and offered to physicians working in the field, to encourage and support them in providing optimized care to their patients. Despite their possible need for revision and improvement, they seem to provide a practical and effective way to promote and consolidate good practice. In addition, they might be used as the basis for refined clinical research.

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Conflict of interest: none declared.

Appendix

Organization of the survey

Expert Committee: Barbara Mulder (Survey chairperson), the Netherlands; Eric Boersma, the Netherlands; Luciano Daliento, Italy; Michael Gatzoulis, UK; Rafael Hirsch, Israel; Harald Kaemmerer, Germany; Folkert Meijboom, the Netherlands; Philip Moons, Belgium; Erwin Oechslin, Switzerland; Jana Popelová, Czech Republic; Erik Thaulow, Norway; Ulf Thilén, Sweden; Jan Tijssen, The Netherlands.

Coordination and Data Management Centre (Euro Heart House, Sophia-Antipolis, France):

KeithMcGregor (ESC Scientific Director); Malika Manini (EHS Operations Manager); Charles Taylor (EHS Database administrator); Claire Bramley, Valérie Laforest (EHS Data Monitors); Susan Del Gaiso (EHS Assistant).

Data Analysis Centre (Cardiology Department, Academic Medical Centre, Amsterdam, the Netherlands): Peter Engelfriet, Jan Tijssen.

National Coordinators: Belgium, Guy De Backer; Switzerland, Peter Buser; Czech Republic, Roman Cerbak; Germany, Uwe Zeymer; Denmark, Per Thayssen; Spain, Angeles Alonso; Finland, Seppo Lehto; France, Jean-Jacques Blanc; UK, Kevin Fox; Greece, Dennis Cokkinos; Hungary, Kristof Karlocai; Israel, Sholmo Behar; Italy, Aldo Maggioni; Lithuania, Virginija Grabauskiene; the Netherlands, Jaap W. Deckers; Poland, Janina Stepinska; Russia, Vyacheslav Mareev; Sweden, Annika Rosengren; Turkey, Tugrul Okay.

There was no national coordinator in the participating countries, which are not mentioned in the above list.

Sponsors: European Society of Cardiology; Dutch Heart Foundation.

Participating Centres and Investigators with numbers of patients included per country:

Armenia (60): Karine Sargsyan, Yerevan. *Austria* (94): Harald Gabriel, Vienna; B. Simma, Michael Fritz, Feldkirch. *Belgium* (170): Werner Budts, Kristien Van Deyk, Philip Moons, Leuven; Julie De Backer, Daniel De Wolf, Johan De Sutter, Bert Suys, Ghent; Martial Massin, Liege; Agnes Pasquet, Marielle Morissens, Brussels; *Czech Republic* (123): Jana Popelova, Ingrid Majerova, Prague; Jindrich Spinar, Anna Necasova, Tomas Brychta, Tomas Zatocil, Brno; *Egypt* (20): Nader Botros, Hala El Farghaly, Ahmed El Maghrabi, Giza; Galal El Said, Sherif El Tobgi, Khalid Sorour, Zeinab Ashour, Howaida G. El Said, Wael Abdelaal, Ali Amer Zakia, Amir Abdulwahab, Khalid Tammam, Cairo; *Estonia* (3): Imbrit Loogna, Tallinn. *France* (46): Laurence Iserin, Paris; Guillaume Jondeau, Boulogne-Billancourt; Yvette Bernard, Besançon. *Germany* (208): Günther Breithardt, Jorge Oberfeld, Thomas Wichter, Stefan Gunia, Muenster; John Hess, Harald Kaemmerer, Annette Wacker, Munich; Reinald Motz, Oldenburg; Peter Lange, Ulrike Bauer, Berlin; Walter Hoffmann, Sabine Nusser, Homburg/Saar. *Greece* (88): Spyridon Rammos, Eftihia Smparouni, Pipina Bonou, Stella Brili, Athens; Periklis Davlouros, Patras; Christos Ntellos, Piraeus. *Hungary* (93): Andras Temesvari, Olga Suranyi, Budapest; Tamas Forster, Marta Hogye, Szeged. *Israel* (234): Rafael Hirsch, Petach Tikva. *Italy* (329): Davide Pacini, Nicola Camurri, Bologna; Luciano Daliento, Padua; Roberto Crepaz, Roberto Cemin, Bolzano. *Lithuania* (99): Alicija Dranenkiene, Lina Gumbiene, Vilnius. *Macedonia* (73): Elizabeta Srbinovska Kostovska, Skopje. *the Netherlands* (706): Chris Jansen, F.J. Meijboom, J.W. Roos-Hesselink, Rotterdam; Harry Crijns, Heidi Fransen, Maastricht; Barbara Mulder, Tanja Megens, Peter Engelfriet, Amsterdam; Arie van Dijk, Colinda Koppelaar, Nijmegen; Hubert Vliegen, Tanja Megens, Leiden; Tieneke Ansink, Lelystad; Dirk Jan van Veldhuisen, T. Steenhuis, Henriette Tebbe, Groningen; Jan Hoorntje, Henriette Tebbe, Annette M. Bootsma, Zwolle; J.B. Winter, H.M.P. Broers, Tilburg; C. Werter, Adrie van den Dool, Roermond; *Norway* (75): Erik Thaulow, J. Westby, Thomas Moller, Oslo; *Poland* (187): Piotr Hoffman, Anna Klisiewicz, Warszawa; Marianna Janion, Marcin Sadowski, Kielce; Olga Trojnarska, Poznan; Tracz Wieslawa, Piotr Podolec, Elzbieta

Suchon, Monika Pieculewicz, Jerzy Sadowski, Krakow; Jan Erecinski, Joanna Kwiatkowska, Gdansk; Jacek Bialkowski, Helena Zakliczynska, Zabrze. *Portugal* (188): Maria Celeste Vagueiro, Claudio Vergulio Antunes David, Lidia de Sousa, Lisbon; Maria da Graça Castro, Coimbra; Vasco Gama Ribeiro, Conceicao Fonseca, V.N. Gaia; Cristina Cruz, Porto. *Romania* (90): Ioan Mircea Coman, Alexandra Gherghina, Mihaela Marin, Bucharest. *Slovakia* (11): Pavol Kunovsky, Viera Vrsanska, Bratislava. *Slovenia* (75): Peter Rakovec, Mirta Kozelj, Katja Prokselj, Ljubljana. *Spain* (317): Jaume Casaldaliga, Joan Cinca, Maite Subirana Domenech, Barcelona; Jose Juan Gomez de Diego, Jose Maria Oliver, Marta Mateos, Madrid; Roberto Barriales-Villa, Pontevedra; Maria Jose Rodriguez Puras, Sevilla. *Sweden* (249): Ulf Naslund, Karin Olsson, Umea; Peter Eriksson, Hrodmar Helgason, M. Dellborg, Gothenburg; Gunilla Forssell, Katarina Ogenholt, Stockholm; Ulf Thilén, Lund. *Switzerland* (309): Peter Eichhron, Schwyz; Heinz Schlapfer, Biel/Bienne; Markus Schwerzmann, Bern; Erwin Oechslin, Zurich; Matthias Pfisterer, Andreas Hoffmann, Basel. *Turkey* (2): Haldun Muderrisoglu, Mehmet E. Korkmaz, Vahide Pimpek, Ankara. *UK* (261): Graham Stuart, Lisa Curthoys, Bristol; G.Y.H. Lip, T. Millane, Deirdre Lane, Ben Sutton, Sara Thorne, Patrick Disney, Birmingham; Michael Gatzoulis, Beatriz Bouzas Zubeldia, London.

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