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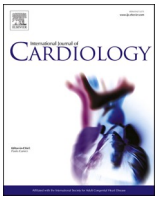

Surveillance of adults with congenital heart disease: Current guidelines and actual clinical practice

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ABSTRACT

Background and aim: Congenital heart disease (CHD) is the most common birth defect with prevalence of 0.8%. Thanks to tremendous progress in medical and surgical practice, nowadays, >90% of children survive into adulthood. Recently European Society of Cardiology (ESC), American College of Cardiology (ACC)/ American Heart Association (AHA) issued guidelines which offer diagnostic and therapeutic recommendations for the different defect categories. However, the type of technical exams and their frequency of follow-up may vary largely between clinicians and centres. We aimed to present an overview of available diagnostic modalities and describe current surveillance practices by cardiologists taking care of adults with CHD (ACHD).

Methods and results: A questionnaire was used to assess the frequency cardiologists treating ACHD for at least one year administrated the most common diagnostic tests for ACHD. The most frequently employed diagnostic modalities were ECG and echocardiography for both mild and moderate/severe CHD. Sixty-seven percent of respondents reported that they routinely address psychosocial well-being.

Conclusion: Differences exist between reported current clinical practice and published guidelines. This is particularly true for the care of patients with mild lesions. In addition, some differences exist between ESC and American guidelines, with more frequent surveillance suggested by the Americans.

1. Introduction

In recent decades, treatment and intervention strategies for patients with congenital heart disease (CHD) have improved and changed their trajectory dramatically. Nowadays, adults outnumber children with CHD and clinicians face an aging population of adults with CHD (ACHD) who continuously increase in numbers and complexity and require specialized cardiac care [1]. Recent European Society of Cardiology

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(ESC) and American College of Cardiology (ACC)/American Heart Association (AHA) guidelines offer diagnostic and therapeutic recommendations for the different defect categories [2,3]. However, the type of technical exams and their frequency of follow-up may vary largely between clinicians and centres, and are based upon historically implemented local hospital protocols, individual experience, and expert opinion. Advice on standardized follow-up intervals would provide further guidance to clinicians, recognizing that these recommendations should be adapted to the individual situation of each patient. A patient's medical and surgical/interventional history, comorbidities, and current clinical health status must be considered, and additional testing may be necessary whenever patients exhibit new symptoms or clinical deterioration. Although many diagnostic modalities are available and potentially beneficial, their advantages and disadvantages must be weighed within the context of available resources (including costs, personnel, and equipment), institutional experience, as well as patient exposure to unpleasant and potentially harmful investigations; it is, thus, important to "choose wisely" [4].

The aims of this paper are to present an overview of available diagnostic modalities and describe current surveillance practices by cardiologists taking care of ACHD patients as obtained through a survey.

2. Current diagnostic modalities

Echocardiography is the first line imaging modality in assessing patients with ACHD, applied routinely in follow-up and in various emergency settings. Echocardiography studies in ACHD are to be supervised and reported by appropriately trained ACHD specialists and lesion-specific protocols have previously been well-described [5–7]. The entire spectrum of echocardiographic modalities (transthoracic, transoesophageal, 3D, contrast, and stress echocardiogram) and sequential segmental analysis are an integral part of cardiac anatomical and functional assessment to ensure that important pathology is not missed. 3D-echocardiography has an established added value in providing "en-face" visualizations of cardiac structures and can help with planning a surgical procedure or a catheter-based procedure. Tissue Doppler imaging and two-dimensional deformation imaging (including global longitudinal strain) have proven to be robust tools and should be integrated into standard clinical practice for longitudinal assessment.

An important, but challenging issue in ACHD is assessment of right ventricle (RV) volumes and function. **Cardiac Magnetic Resonance Imaging (CMRI)** is the preferred and the reference technique to evaluate the RV for those; however when not possible, follow-up of RV size and function may be achieved through echocardiography, including 3D-echo, in expert centres [6–10]. Indirect measures of RV systolic function, such as fractional area change and tricuspid annular plane systolic excursion from a four-chamber view, are also widely used and have shown prognostic value. 2D speckle tracking-derived strain of the RV free wall, appears to be feasible and reproducible. Changes in ventricular dimensions and systolic function over time may be helpful to guide therapy decisions [11].

CMRI is ideal for accurate quantification of ventricular volumes, ejection fraction (EF), valvular regurgitation, calculation of pulmonary and systemic blood flow, and myocardial fibrosis assessment [7]. CMRI facilitates 3D anatomical reconstruction, is not restricted by body size or acoustic windows, and has rapidly improving spatial and temporal resolution. Furthermore, the lack of radiation makes it a useful diagnostic tool when serial evaluations are needed (for routine follow-up of aortic coarctation or to monitor aortic dimensions, RV volumes and ejection fraction in tetralogy of Fallot, congenitally corrected transposition of the great arteries, or after atrial switch procedure). CMRI allows calculation of systemic and pulmonary blood flow in patients with multiple sources of blood supply and therefore of pulmonary vascular resistance, when combined with invasive catheterization. In patients with complex single ventricle circulation such as Fontan physiology, flow analysis provides accurate non-invasive haemodynamic data, including collateral flow and shunt through a fenestration if present. Tissue characterisation for myocardial fibrosis is a unique capability of CMRI. Late gadolinium enhancement CMRI for focal fibrosis and interstitial fibrosis T1 mapping imaging are increasingly being applied in ACHD for their diagnostic and potential prognostic value. ACHD patients with conventional pacemakers or ICDs can undergo CMRI within guidelines where local support is available [12]. 3D-CMRI imaging can be integrated into electrophysiology procedures to inform and guide therapy.

3-dimensional Cardiac Computer Tomography (CCT) and CMRI reconstructions are both applied for virtual reality planning, for assessing of intervention, for patient-specific 3D prints, or for routine follow-up in selected cases. CCT has high spatial resolution and short acquisition time and is particularly relevant for imaging the great vessels, coronary and collateral arteries, and for parenchymal lung disease. Recent rapid developments have substantially reduced the amount of radiation, achieving <5mSV for a combined CCT coronary, pulmonary, and aortic angiogram. This made CCT more attractive than previously in ACHD patients for specific indications such as coronary artery pathology and/or detailed assessment of collaterals [13]. CCT is particularly useful in emergency settings, where it may have advantages over echocardiography and CMRI due to being less susceptible to prosthetic valve artefacts. ACHD is pre-eminently a sub discipline in cardiology where multimodality imaging is applicable. The diagnostic utility of a multimodality approach is greater than the sum of individual tests. Therefore, recommendations for imaging approaches for surveillance of ACHD patients highlight the potential of a multimodality approach for an optimal balance between utilization of tests and diagnostic yield [6].

***Cardiopulmonary exercise testing (*CPET)** is an objective, reproducible, and valuable clinical tool for an integrative assessment of the pulmonary, cardiovascular, and musculoskeletal system. CPET provides a more comprehensive assessment, as it provides more information, to the standard exercise testing, and is the preferred method to evaluate exercise capacity in ACHD.

Many ACHD patients have learned to adjust to a lower exercise tolerance since childhood: they often overestimate their exercise capacity and may not perceive a slow, gradual decline [14]. A discrepancy between perceived exercise tolerance and objectively assessed exercise capacity is very common even in 'asymptomatic' patients [15]. Reference values from healthy volunteers thus cannot be applied to ACHD patients with a wide anatomic and pathophysiologic spectrum. Sex- and age-specific and lesion specific reference values for exercise capacity in ACHD are essential [16]. Formal exercise capacity has been linked with self-reported physical functioning [17]. CPET is helpful not only with the assessment of functional capacity, but also offers prognostic information in identifying patients at risk for morbidity and mortality by an integrative assessment of exercise capacity, chronotropic competence, heart rate recovery, ventilatory efficiency, blood pressure response, desaturation, and arrhythmias [15,18–24]. Serial testing is essential to unmask subclinical changes which may prompt consideration for interventions. The **6-min walk test (6MWT)** is an alternative for cyanotic patients, disabled patients, or patients who cannot tolerate wearing a mask. Information provided by the 6MWT is limited, however serial tests do have prognostic value [25,26].

Although guidelines do not recommend the routine use of **catheterisation**, as this is an invasive diagnostic procedure, it may be indicated in specific circumstances. It provides information on ventricular and valvular function and hemodynamic. Examples include patients with (suspicion of) pulmonary arterial hypertension and those for whom other diagnostic techniques fail to provide sufficient information. It is important to remember to perform cardiac catheterisation in Fontan patients at low threshold as symptoms occurs [27].

Currently, there is increasing evidence of the diagnostic and prognostic value of different classes of **laboratory markers.** Although their use in ACHD is challenging due to cut-off variability among different CHD lesions, neuro-hormones such as natriuretic peptides (BNP) and N-terminal-pro-BNP (NT-proBNP) or markers of myocardial injury (high-

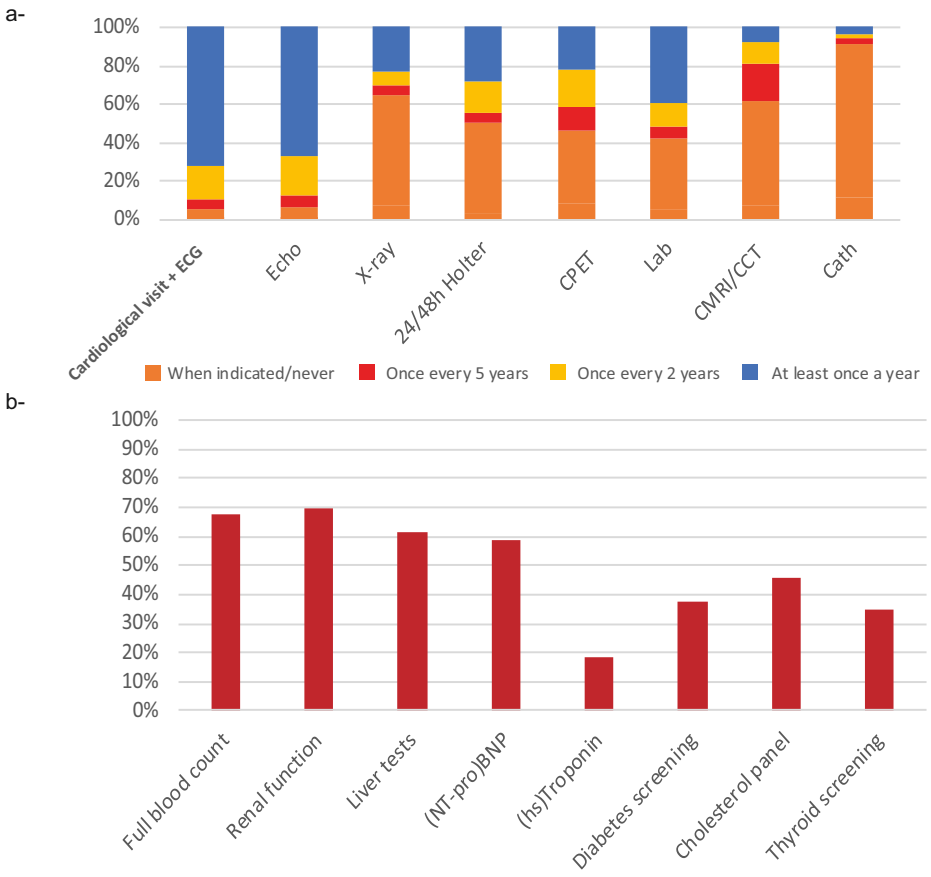

Fig. 1. (a) Frequency of prescription: (a) diagnostic tests, (b) laboratory tests CPET: Cardiopulmonary exercise test, CMRI: Cardiac Magnetic Resonance Imaging, CCT: Cardiovascular Computer Tomography.

sensitivity troponins; hs-TNT) or inflammation (high-sensitivity C-reactive protein; hs-CRP) have an established role in routine care as they relate to cardiac function, exercise capacity, cardiovascular events and mortality [28–38]. Serial NT-proBNP measurements increase the prognostic value of the periodic ACHD follow-up assessment [32]. Further, elevations in hs-TNT and hs-CRP relate to been linked with worse outcome in ACHD patients [29,30,33,35,36,39]. Other markers may also be useful in specific situations, for instance measuring hemoglobin, hematocrit and iron status in cyanotic patients or albumin, alfa-fetoprotein, and liver function in Fontan patients.

A resting **12-lead ECG** and measuring **transcutaneous oxygen saturation** should be part of every cardiological consultation in ACHD. The detection rate on **24/48-h Holter recording**, particularly for paroxysmal or only sporadic arrhythmia, is limited due to the short duration of the recordings. An alternative to detect paroxysmal arrhythmias is a **patient-activated device** that allows rhythm recording each time that the patient experiences a symptomatic event. **Implantable loop recorders** are a further option to detect sporadic arrhythmias and are primarily indicated in patients who experience syncope.

2.1. Psychosocial assessment

In addition to a proactive approach to monitoring and optimizing physical health, strategies to identify and manage psychological distress represent an important component of comprehensive ACHD care. Up to half of ACHD patients will experience a mood and/or anxiety disorder during their lifetime [40]. Psychological distress in ACHD patients is undertreated, and associated with worse functional class, higher mortality, and more healthcare utilization [41–48]. Identification of significant psychological distress can be addressed through routine screening or dialogue during clinical visits. Psychological screening using surveys has been recommended for ACHD patients, although the clinical effectiveness of this practice is largely unknown and requires dedicated personnel and other resources [49,50]. Screening surveys have the advantage of quantifying psychological distress over time, although concerns include the potential for false negatives/positives and allocation of mental health resources to screening rather than treatment [51,52]. An alternative strategy is to initiate dialogue about psychological well-being during the visit, and document this in clinic notes. This approach conveys clinicians' empathy regarding challenges of living with CHD and may identify patients who would benefit from referral to mental healthcare.

3. Survey of current surveillance practices

3.1. Methods

To investigate current practices with regards to diagnostic testing and follow-up in ACHD, the writing group, developed a questionnaire between May and December 2021 in collaboration with the ESC marketing department. The writing group aimed for a reasonable balance between comprehensiveness and time burden for completion. The final questionnaire included items about the 15 most frequent ACHD diagnoses, categorized according to the ESC classification as mild, moderate, or severe disease [27].

For each diagnosis, we asked multiple choice questions about the frequency the cardiologists administrated the following diagnostic tests: cardiology clinic visit plus ECG, echocardiogram, chest X-ray, 24/48-h Holter monitoring, CPET, laboratory investigations, CMRI/CCT, and cardiac catheterization. The following were response options: whenever

Fig. 2. Frequency of prescription of diagnostic tests per CHD diagnosis (a) Cardiological visit + ECG (b) Echocardiography, (c) X-ray, (d) 24/48 h Holter monitor, (e) Exercise testing, (f) Laboratory investigations, (h) MRI or CT, (g) Cardiac catherisation.

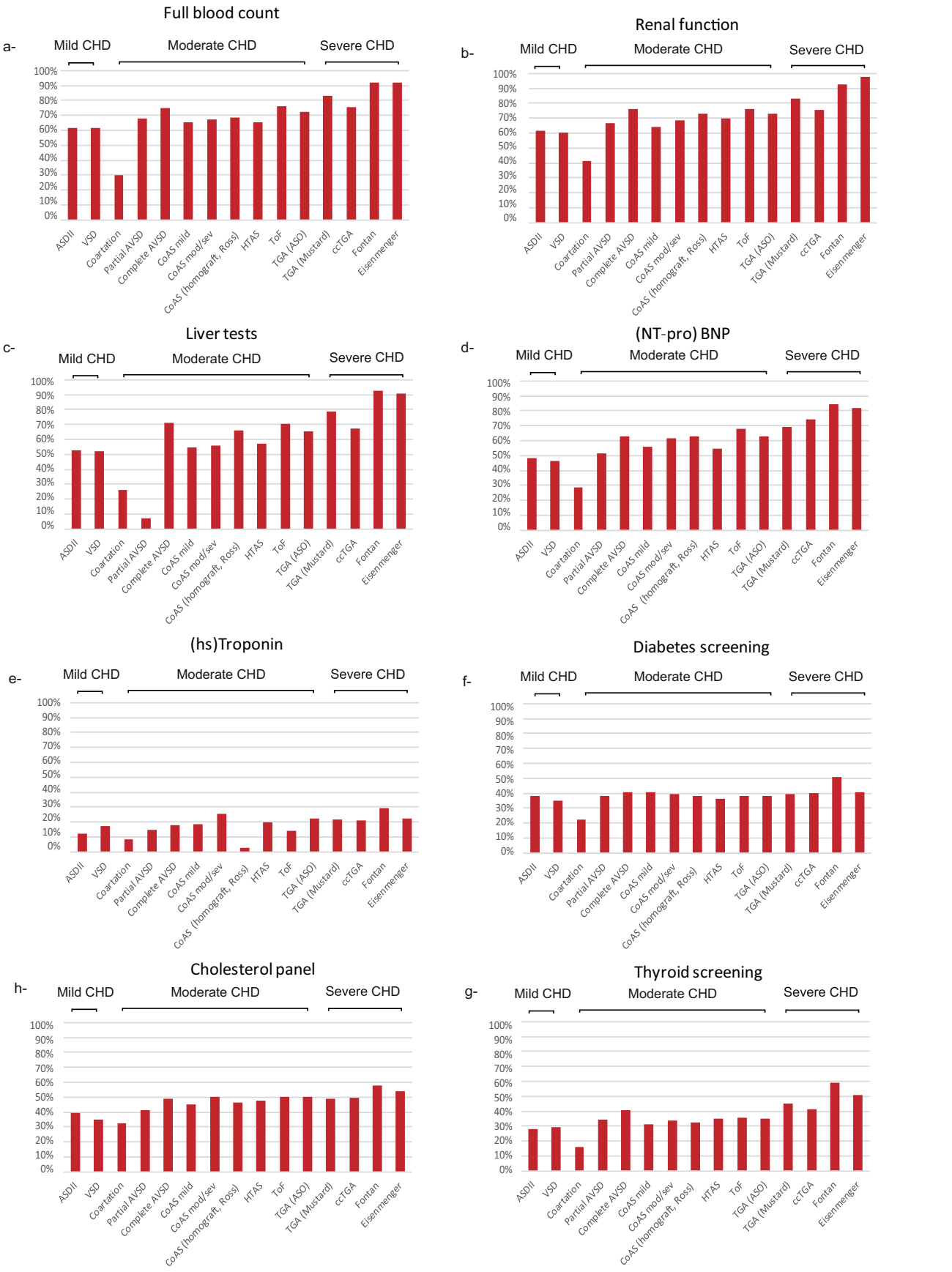

Fig. 3. Frequency of prescription of laboratory tests per CHD diangosis: (a) full blood count, (b) renal function, (c) liver tests, (d) (NT-pro) BNP, (e) (hs) Troponin, (f) diabetes screening, (h) cholesterol panel, (g) thyroid screening.

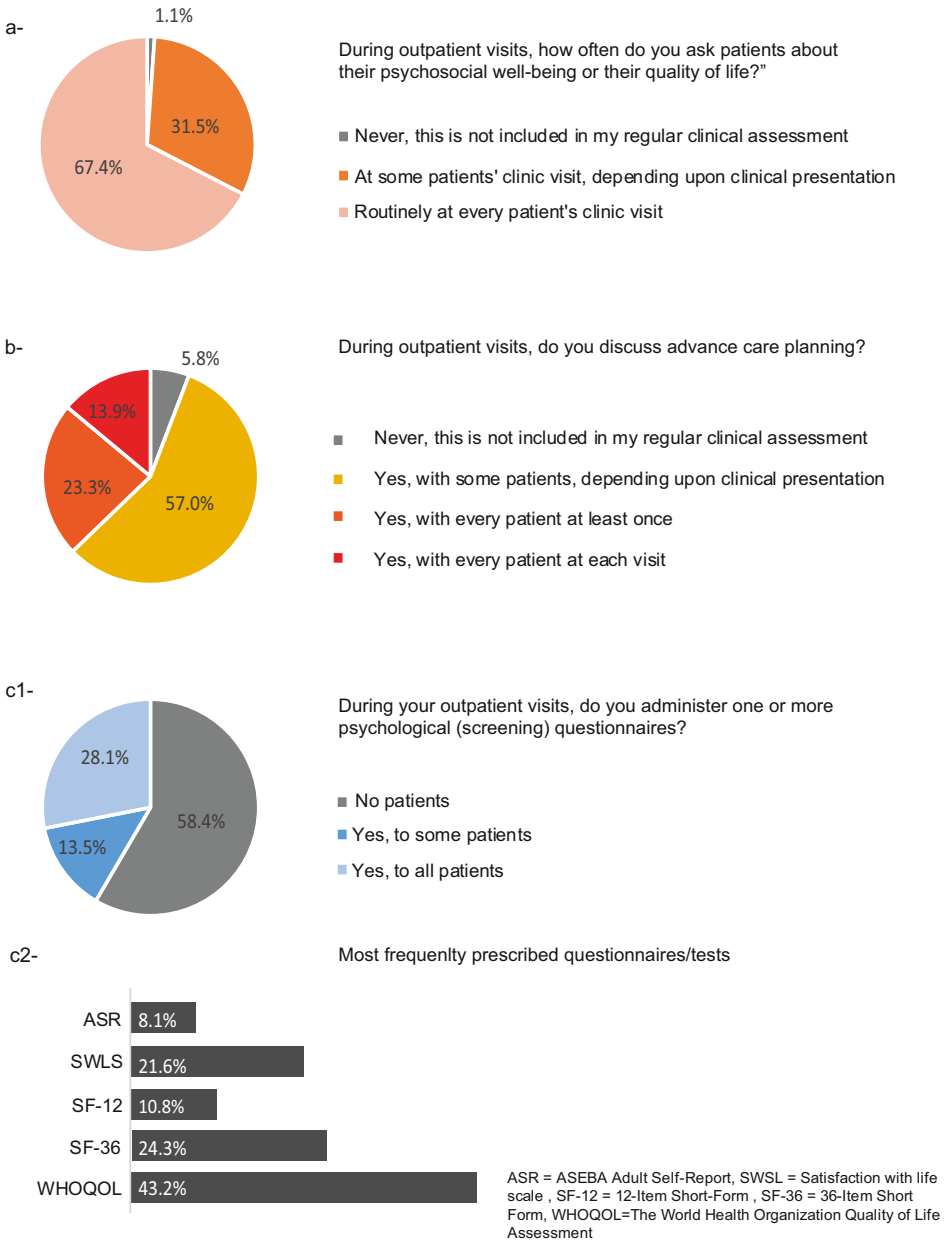

Fig. 4. Investigations regarding psychosocial well-being and quality of life.

indicated, once every 5 years, once every 2 years, or at least once a year. In addition, 3 questions regarding psychosocial assessment were included.

On January 31st, 2022, a link to a website with the questionnaire was distributed to cardiologists, inviting participation from physicians with at least one year of ACHD experience. The website remained open for one month.

Categorical data are reported in percentages. Statistical analysis of the data was processed using IBM Statistics SPSS v28.0 version for Windows.

4. Results

The survey was sent to an estimated 1200 cardiologists with at least one year of ACHD experience, of whom 89 completed the survey. Findings are reported in Figs. 1–4. The most frequently employed diagnostic modalities were ECG and echocardiography. Most respondents performed an ECG at least once a year in all patients with moderate or severe CHD (Fig. 2a), and reported a lower frequency (once every 2 or 5 years) for patients with mild CHD. Most cardiologists performed an echocardiogram at every visit (Fig. 1a).

Sixty-seven percent of respondents reported that they routinely address aspects of psychosocial well-being during clinic visits with all patients (Fig. 4). Further, 23% of respondents reported that they discuss advance care planning with every patient, and 57% of them discuss this topic only on indication.

Table 1 presents an overview, according to defect subtype, of cardiac diagnostic testing as performed currently in clinical practice (survey results) in comparison to the most recent ESC and ACC/ AHA guidelines [2,27,53].

5. Discussion

This study revealed important discrepancies between clinical diagnostic practice and European and American ACHD guidelines, particularly for the surveillance of patients with mild CHD. Specifically,

Table 1
Diagnostic testing in ACHD according to the questionnaire, ESC and ACC/AHA guidelines.

CHD	Cardiological visit ECG			Echo			Other diagnostic tests		
	Survey	ESC	AHA	Survey	ESC	AHA	Survey	ESC	AHA
ASD II*	Every 5 years	Not indicated	Every 3–5 year	Every 5 years	Not indicated	Every 3–5 year			
VSD*	Every 5 years	Every 5 years	Every 3 year	Every 5 years	Not indicated	Every 3 year			
Aortic Coarctation	Every year	Every year	Every 2 years	Every 1–2 years	Not indicated	Every 2 years	CCT/CMRI (every 1–2 years)	CMRI (every 3-5 years)	CCT/CMRI (every 3-5 years), exercise testing (every 3 years)
Partial AVSD	Every year	Every 3–5 years	Every 2–3 years	Every 1–2 years	Every 2–3 years	Every 2–3 years			
Complete AVSD	Every year	Every 2–3 years	Every 2–3 years	Every year	Every 2–3 years	Every 2–3 years	Lab (routinely)		
Congenital AoS** (mild V max <3 m/s)	Every year	Regular (on individual basis)	Every 2 years	Every 1–2 years	Regular (on individual basis)	Every 3–5 year			CCT/CMRI (Ao dilatation)
Congenital AoS ** (moderate/ severe, Vmax>3 m/s)	Every year	Regular (on individual basis)	Every 2 years	Every year	Regular (on individual basis)	Every 1–2 years (moderate), every 12–6 mo (severe)	Exercise test (routinely) Lab (routinely)	CCT/CMRI (for restenosis/ aneurism)	CCT/CMRI (Ao dilatation)
Congenital AoS valvular (after homograft/Ross operation)	Every year	Regular	Every 2 years	Every year	Regular (valve and aortic root)	Every 3–5 years (mild), Every 1–2 years (moderate), every 12–6 mo (severe)		CCT/CMRI (regularly)	
HTAD or BAV (aorta diameter < 45 mm)	Every year	Every year	Regular	Every year	Regular	Regular[$]	CCT/CMRI (every 1–2 years)	CCT/CMRI (regularly)	BAV: CCT/CMRI (regularly) Marfan: imaging surveillance (every 3–5 years)[$]
ToF***	Every year	Every year	Every 1–2 year	Every year	Every year	Every 2 years	CCT/CMRI (every 5 years) Holter (every 1 or 2 years) Exercise test (every 1–2 years) Lab (every 1–2 year)	CMR (regularly)	CCT/CMRI (every 3–5 years), exercise testing (every 3 years)
TGA (arterial switch)	Every year	Every year	Every 1–2 year	Every year	Every year	Every 1–2 years	Exercise test (every 1–2 years) Lab (every 1–2 year)		CCT/CMRI (every 3–5 years), exercise testing (every 3–5 years)
TGA (Mustard/ Senning)	Every year	Every year	Every year	Every year	Every year	Every 1–2 years	Holter (every 1–2 years) Exercise test (every 1–2 years) Lab (every 1–2 year)		CCT/CMRI (every 2–3 years), exercise testing (every 3 years) Holter (every 2 years) Pulse oximetry (every year)
ccTGA	Every year	Every year	Every year	Every year	Every year	Every 1–2 years	Holter (every 1–2 years) Exercise test (every 1–2 years) Lab (every 1–2 year)		CCT/CMRI (every 3–5 years), exercise testing (every 3–5 years) Holter (every 1–5 years)
Fontan	Every year	Every year	Every year	Every year	Every year	Every year	CCT/CMRI (every 5 years) Holter (every 1–2 years) Lab (every year)	Blood test (every year), Exercise test (every year), CMR and hepatic assessment (on individual basis)	CCT/CMRI (every 3 years), exercise testing (every 3 years) Holter (every year) Pulse oximetry (every year)
Eisenmenger Syndrome	Every year	Every year (every 6 mo)	Every year (every 6 mo)	Every year		Every year	Exercise test (routinely) Lab (every year)		Pulse oximetry (every visit) Exercise test (every year/every 6 mo)

*Small, untreated OR after closure in the past, without PH, ** Valvular or sub-valvular, unrepaired, ***after repair in childhood, $ **BAV Aorta diameter (root or ascending aorta or both) ≥ 40 mm:** Regular imaging (Echo, CT or MRI) follow-up dependent of the rate of growth.
Marfan: - Stable aortic diameter: Annual surveillance with Echo.
- Aortic root replacement: If normal and unchanged for 2 years, then surveillance every 2 years.
ASD II = Atrial Septal Defect, type II, VSD = Ventricular Septal Defect, AVSD = Atrioventricular Septal Defect, AoS = Aortic Stenosis, HTAD = Hereditable Thoracic Aortic Disease, BAV = Bicuspid Aortic Valve, ToF = Tetralogy of Fallot, TGA = Transposition of the Great Arteries, ccTGA: congenitally corrected Transposition of the Great Arteries.
ECG = Electrocardiogram, Echo = Echocardiography, CPET = Cardiopulmonary Exercise Test, CMRI = Cardiac Magnetic Resonance Imaging, CT = Computer Tomography, Lab = Laboratory Investigations, Cath = Cardiac Catheterization.

respondents administer diagnostic testing for patients with less severe CHD more frequently than recommended by guidelines, particularly for patients with moderate/severe congenital aortic stenosis, post Ross procedure and atrioventricular septal defects.

Regarding other diagnostic tests, 10% of the respondents routinely perform cardiac catheterization. While this is justifiable in specific cases such as in symptomatic Fontan patients, guidelines discourage its use with stable ACHD patients and instead advise that it should be reserved for addressing specific clinical questions. Therefore, it seems that cardiac catheterization is used too often, especially since it is an invasive procedure. In addition, the use of laboratory testing was more frequently reported among respondents than recommended in the guidelines. This may be the result of a more liberal use of biomarkers in recent years, especially NT-proBNP, which has proven useful especially in patients with moderate or severe CHD [32]. It might be that these studies will change future guidelines. The practice of performing echocardiograms at every follow-up visit may be questionable and perhaps unnecessary. This implementation gap between guidelines and actual clinical practice reflects potentially unjustifiable visits and tests with a negative impact on limited resources, additional burden on the healthcare system and on the already high costs. The concept of"choosing wisely" can support the selection of tests and treatments in the best interest of individual patients, while also avoiding overuse of diagnostic procedures, some of which carry potential patient harm [4,54]. The overutilization of diagnostic methods may be due to various underlying factors. One potential driver could be the apprehension among physicians regarding the risk of underdiagnoses, fear of malpractice and sense of medical obligation. Additionally, pressure from patients and patient-doctor relationship may play a role as well [55].

In contrast to the potential over-use of certain diagnostic modalities, the survey revealed under-use (compared to guidelines) of CPET, which has demonstrated effectiveness to detect asymptomatic reduction in exercise capacity and to document changes in aerobic capacity over time. The reduction or decline of objective exercise capacity is important as this can be taken into account when deciding on the indication for preventive therapy.

Further, best practices for routine screening of ACHD patients for risk factors for acquired cardiovascular disease (e.g.: cholesterol, blood sugar), are currently unknown. Indeed, more attention is warranted for prevention of atherosclerotic heart disease in this young population [54].

Moreover, we recommend that new guidelines include advice on the level of evidence supporting the recommended follow-up intervals for various tests, as detailed information on this aspect is currently lacking in the existing guidelines. Both ESC and AHA lack of specific guidelines for psychosocial assessment in ACHD. Nevertheless, the ESC guidelines designate psychologists as essential specialists for the ACHD treating team. Consequently, there is a need for the formulation of detailed recommendations regarding psychosocial assessment in future guidelines.

In our study, two-thirds of interviewed cardiologists routinely inquire about the quality of life of their ACHD patients during every visit. Systematic documentation of these data in clinical files can ensure referrals to mental health services if necessary, ensuring prompt intervention and support for patients' psychological well-being.

5.1. Survey limitations

Study limitations include the low response rate (although the denominator of cardiologists with at least one year of ACHD experience who received the survey was estimated) as well as generalizability because we did not collect information regarding demographic, gender, or practice characteristics of survey respondents.

6. Conclusions

In conclusion, differences exist between reported current clinical practice and published guidelines. This is particularly true for the care of patients with mild lesions, for whom the standard frequency of follow-up in current practice may be unnecessarily high. In addition, some differences exist between ESC and American guidelines, with more frequent surveillance suggested by the Americans.

CRediT authorship contribution statement

Jolien W. Roos-Hesselink: Writing – review & editing, Writing – original draft, Validation, Supervision, Project administration, Methodology, Data curation, Conceptualization. **Chiara Pelosi:** Writing – review & editing, Writing – original draft, Visualization, Methodology, Formal analysis, Data curation. **Margarita Brida:** Writing – review & editing, Writing – original draft, Validation, Methodology, Conceptualization. **Julie De Backer:** Writing – review & editing, Writing – original draft, Methodology, Conceptualization. **Sabine Ernst:** Writing – review & editing, Writing – original draft. **Werner Budts:** Writing – original draft, Writing – review & editing. **Helmut Baumgartner:** Conceptualization, Methodology, Validation, Writing – original draft, Writing – review & editing. **Erwin Oechslin:** Methodology, Writing – original draft, Writing – review & editing. **Daniel Tobler:** Writing – original draft, Writing – review & editing. **Adrienne H. Kovacs:** Conceptualization, Methodology, Writing – original draft, Writing – review & editing. **Giovanni Di Salvo:** Writing – review & editing. **Jolanda Kluin:** Writing – review & editing. **Michael A. Gatzoulis:** Writing – original draft, Writing – review & editing. **Gerhard P. Diller:** Conceptualization, Methodology, Supervision, Validation, Writing – original draft, Writing – review & editing.

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