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Standardization in paediatric echocardiographic reporting and critical interpretation of measurements, functional parameters, and prediction scores: a clinical consensus statement of the European Association of Cardiovascular Imaging of the European Society of Cardiology and the Association for European Paediatric and Congenital Cardiology

Massimiliano Cantinotti [1]*, Giovanni Di Salvo [2], Inga Voges[3], Francesca Raimondi[4], Gerald Greil[5], Almudena Ortiz Garrido[6], Tara Bharucha[7], Heynric B. Grotenhuis[8], Martin Köstenberger[9], Beatrice Bonnello[10], Owen Miller[11], and Colin J. McMahon[12,13,14]

[1]Department of Pediatric Cardiology and Congenital Heart Disease, National Research Council-Tuscany Region G. Monasterio Foundation (FTGM), Massa, Pisa 54100, Italy; [2]Paediatric Cardiology and Congenital Heart Disease, Woman and Children's Health Department, University of Padua; Experimental Cardiology, Paediatric Research Institute (IRP), Padua, Italy; [3]Department of Congenital Heart Disease and Pediatric Cardiology, DZHK (German Centre for Cardiovascular Research), Partner Site Hamburg/Kiel/Lübeck, University Hospital Schleswig-Holstein, Kiel, Germany; [4]Division of Pediatric Cardiology, ASST Giovanni XXIII, Bergamo, Italy; [5]Division Pediatric Cardiology, UT Southwestern, Dallas, TX, USA; [6]Department Pediatric Cardiology, Malaga, Hospital Materno Infantil, Andalucia, Spain; [7]Department of Paediatric Cardiology, University Hospital Southampton, Southampton, UK; [8]Department Pediatric Cardiology, Wilhelmina Children's Hospital/UMCU, Utrecht, The Netherlands; [9]Department of Pediatrics, Division of Pediatric Cardiology, Medical University of Gratz, Gratz, Austria; [10]Great Ormond Street Hospital for Children, London, UK; [11]Department Pediatric Cardiology, Evelina London Children's Hospital, London, UK; [12]Department Paediatric Cardiology, Children's Health Ireland at Crumlin, Dublin, Ireland; [13]School of Medicine, University College Dublin, Belfield, Dublin, Ireland; and [14]Maastricht School of Health Professions Education, Maastricht, The Netherlands

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This document has been developed to provide a guide for basic and advanced reporting in paediatric echocardiography. Furthermore, it aims to help clinicians in the interpretation of echocardiographic measurements and functional data for estimating the severity of disease in different paediatric age groups. The following topics will be reviewed and discussed in the present document: (i) the general principle in constructing a paediatric echocardiographic report, (ii) the basic elements to be included, and (iii) the potential and limitation of currently employed tools used for disease severity quantification during paediatric reporting. A guide for the interpretation of Z-scores will be provided. Use and interpretation of parameters employed for quantification of ventricular systolic function will be discussed. Difficulties in the adoption of adult parameters for the study of diastolic function and valve defects at different ages and pressure and loading conditions will be outlined, with pitfalls for the assessment listed. A guide for careful use of prediction scores for complex congenital heart disease will be provided. Examples of basic and advanced (disease-specific) formats for reporting in paediatric echocardiography will be provided. This document should serve as a comprehensive guide to (i) structure a comprehensive paediatric echocardiographic report; (ii) identify the basic morphological details, measures, and functional parameters to be included during echocardiographic reporting; and (iii) correctly interpret measurements and functional data for estimating disease severity.

Keywords echocardiography • children • congenital heart disease • reporting • formats • scores

* Corresponding author. E-mail: cantinotti@ftgm.it

Introduction

Guidelines and standards for paediatric echocardiography have been well published over the past decade,[1–12] including recommendations for a standard protocol,[1] quantification,[7] structured reporting,[2,4] and training.[5] These documents[2,4,5,7] address the role of echocardiography in different conditions, from normal to different pathological states. Recommendations for the content of structured reporting systems have also been published,[2,13,14] yet no established standard reporting formats are available to facilitate the following objectives: (i) to promote standardized care, (ii) to avoid inadvertent exclusion of potentially relevant information, (iii) to distinguish among essential and optional elements, and (iv) to optimize time use.[13,14]

Despite the availability of recommendations[7] for quantification methods during the performance of a paediatric echocardiogram and international nomenclature for congenital heart disease (CHD),[14–17] estimation of disease severity often remains a challenge. The use of Z-scores[18–28] has been accepted for two-dimensional (2D) measures, and robust normal value sources are currently available.[18–21] Which of the available Z-score sources should be adopted remains however unclear.[18–21] Even the evaluation of ventricular sizes in children[29–38] is not so standardized as in adults.[33] The greatest difficulties however are encountered for the quantification of functional data. Many aspects of Doppler[18,27,28] and colour Doppler flow evaluation[39–46] in the paediatric age group have not yet been completely defined. Significant uncertainty exists in the evaluation of diastolic function, as patterns of altered diastolic function at different ages, and in different loading and pressure conditions, have been poorly defined.[47–63] Similarly, quantification of valvular defect severity furthermore may be troublesome since the adult parameters to estimate valvular defects[29,45,46] have not been completely validated in the paediatric cohort.[64–96] Also, despite septal defects being very common in the children, echocardiographic parameters to grade shunt size are not completely defined.[39–46] The introduction of new three-dimensional (3D)[29,31,32] and speckle tracking echocardiography (STE) modalities[97–108] poses new challenges in interpretation of results. Lastly, there is a series of echocardiographic parameters and prediction scores for biventricular risk estimation in complex CHD,[109–128] which remain contentious. Other scores have also been poorly applied so far [such as those for prediction of aortic coarctation (CoA)].[129–134]

The aims of this manuscript are:

(i) To evaluate the strengths and limitations of published recommendations for reporting paediatric echocardiography with CHD
(ii) To discuss which elements should be required, rather than optional, in a standard report
(iii) To propose a uniform standard for reporting in paediatric echocardiography
(iv) To review the potential and limitation of tools currently employed for disease severity quantification during paediatric reporting, including Z-scores; parameters for classifying ventricular systolic and diastolic function, valvular defects, and shunt lesions; and which scores should be used for complex CHD

This document has been written by the members of the Imaging Working Group of the Association for European Paediatric and Congenital Cardiology and by the members of the Grown-Up Congenital Heart Disease Taskforce of the European Association of Cardiovascular Imaging of the European Society of Cardiology (ESC). The document follows criteria for the expert consensus paper of the ESC.[135] Categories indicated in the ESC Clinical Consensus Statement have been used for clinical advice.[136]

Table 1 Principles of structured, balance, and practical reporting

Consistency	There should be an organized structure, fixed elements, and defined terminology
Flexibility	Addition of elements and free text should be allowed
Completeness	Allow inclusion of all potentially relevant information
Conciseness	Easy to understand, fast to read
Reproducibility	Adequate for various settings, diseases, and operator's skill
Practical	Easy to apply, pertinent to daily issues
Able to evolve over time	Compatible with evolution in knowledge, advances of new techniques
Digital and compatible	Allow interoperability between electronic medical record systems

General principles in the building of a report

The construction of a structured report requires consistency with regard to (i) a standardized protocol of imaging acquisition with a fixed order of items to be followed, (ii) essential elements to be included, (iii) availability of a solid nomenclature of definitions, and (iv) classification of CHD and grading of their severity.[2] The report should be flexible (with the possibility to add or subtract elements and free text spaces), complete, concise, reproducible (for different settings, diseases, and operators' skill), practical, capable of evolving over time, and applicable between different electronic medical record systems[4,5] (*Table 1*).

Consistency

The order to be followed and essential elements to be included: the segmental approach

The segmental approach is recognized as the gold standard in evaluation of CHD.[1,4–6] This detailed anatomical and functional analysis, including the views and the required projections and their sequential order, and the essential elements to evaluate have been well defined by standardized review.[1,4,7–12]

The use of a common nomenclature

Over the years, various consensus papers[15–17] have tried to establish an international system for classification and coding of different CHD, as well as of their surgical/interventional treatment. Paediatric echocardiographic systems to grade the severity of many congenital and acquired defects however remain limited yet.[1–4,7–12]

Clinical Advice:

Strength of advice	A standardized protocol of imaging acquisition (e.g., the segmental approach) is advised for imaging acquisition in paediatric echocardiography.	
	The use of an international system for the classification and coding of different CHD is advised for the reporting of paediatric echocardiography.	
	Due to variability of the code for CHD classification, the same code/classification of CHD in the same institution and between different practice groups (e.g., children and adult congenital heart disease) in the institution is advised.	

Gap in knowledge:

- System and code for CHD should be uniformed.
- Systems to classify disease severity need to be implemented.

Flexibility

A single report for all CHD or a report for every single CHD?

Due to the large variety of congenital and acquired cardiac lesions presenting in the paediatric age group, reports will vary extensively for different defects.[5] Whether a unique, flexible report for all cardiac lesions or single reports for specific lesions should be employed is debatable. The use of a single format for all cardiac lesions requires flexibility, particularly in terms of adding or removing elements.[3] Thus, in a basic format, various elements can be added, whereas in a very complex format, unneeded information can be removed. Examples of basic and complete formats for reporting are provided in *Tables 2–5* and Supplementary data online, *Table S1*. The use of separate formats for various cardiac lesions would require the availability of multiple formats, due to the great variety of CHDs. The use of multiple formats may be helpful for standardization in reporting and serve as a guide for the clinician, especially younger trainees, in the diagnosis of complex CHDs. The CHD-specific format should indicate all the relevant details that are required for a specific cardiac lesion. The major issue related to multiple formats is represented by complex and rare CHDs and by the presence of associated CHD.[1–12] Thus, even lesion-specific formats may prove inadequate for the complete evaluation of difficult anatomy; therefore, flexibility is required. As for single formats, in complex CHD-specific formats, unnecessary information, or data that has not been acquired for technical reasons, may need to be removed. It is important to note that not all the measurements are required at each examination.[7]

In Supplementary data online, *Tables S2–S13*, reporting formats for major groups of CHDs before and after surgical and/or percutaneous correction/palliation are presented. It starts from simple defects such as left-to-right shunt [septal defects and patent arterial duct (PDA)] (see Supplementary data online, *Tables S2–S4*] to a more complex CHD [e.g. atrioventricular septal defects (AVSD), transposition of the great arteries, anomalous pulmonary venous return, and conotruncal defects; see Supplementary data online, *Tables S5–S11*). Complex univentricular CHDs before (see Supplementary data online, *Table S12*) and after (see Supplementary data online, *Table S13*) different stages of Fontan palliation have also been presented. Lastly, examples of rare CHDs such as congenital mitral stenosis and aorto-pulmonary are provided (see Supplementary data online, *Table S14*).

Clinical Advice:

Strength of advice		
	The use of CHDs specific format may provide advantages over the use of single formats in pre-operative/interventional examinations, promoting standardization, serving as a guide for reporting, and helping to collect all relevant information for a single disease.	
	The use of single formats may be acceptable for normal or quick post-operative/interventional follow-up echocardiographic examination.	
	Flexibility is required both in single and in lesion specific formats.	

Gap in knowledge:

An analysis of the advantages and limitations in different methods of reporting has not yet been performed.

Table 2 Example of basic 2D and Doppler assessment format for reporting a normal paediatric echocardiographic examination

Patient name:
Date of birth (DOB):
Date of examination:
Age (years/months):
Weight (kg): Length (cm): BSA (m^2):
Arterial pressure (mm Hg): HR: b.p.m.
Oxygen saturation: % Rhythm:
Department: Operator:

Situs:	Solitus
Position within the chest:	Levocardia
AV connection	Concordant
Ventriculo-arterial connection:	Concordant
Pulmonary venous return:	Normal
Systemic venous return	Normal
Inter-atrial septum	Intact
Interventricular septum	Intact
Cardiac chambers	
RA	Normal dimensions
LA	Normal dimensions
LV	Normal dimensions, volumes, and thickness. Normal systolic and diastolic functions
RV	Normal dimensions, volumes, and thickness. Normal systolic and diastolic functions
Valves	
TV	Physiologic insufficiency RV–RA difference of pressure: mm Hg
MV	Normal anatomy, no insufficiency, no stenosis
Aortic valve	Normal anatomy, no insufficiency, no stenosis Aortic sinuses and ascending aorta of normal dimensions
Pulmonary valve	Normal anatomy, physiologic insufficiency, no stenosis
Great vessels	
Main pulmonary artery	Normal dimension
Right pulmonary artery:	Normal dimension
Left pulmonary artery:	Normal dimension
Aortic arch	Normal anatomy and normal vessels take-off
Arterial duct/collaterals	Absent
Coronary arteries	Normal origin and dimension
Pericardium	No effusion
Others	
Quality of the examination	
Conclusions	No evidence of structural and functional heart disease. Findings within the range of normality for the age
Signature:	

BSA, body surface area; b.p.m., beats for minute.

Table 3 A complete 2D and Doppler assessment format for reporting in paediatric echocardiography with basic anatomical and functional detail, basic and advanced measurements, and functional parameters

Patient name
Date of birth:
Date of examination:
Age (years/months):
Weight (kg): Length (cm): BSA (m^2):
Arterial pressure (mm Hg): HR: b.p.m.
Oxygen saturation: % Rhythm:
Department: Operator:
Echo machine: Software employed for 3D, strain analysis:

	Basic anatomical/functional detail	Basic measures/functional parameters	Advanced measures/functional parameters
Situs:	Solitus:		
	Ambiguus:		
	Inversus:		
Position within the chest:	Levocardia:		
	Mesocardia:		
	Dextrocardia:		
AV connection	Concordant:		
	Discordant:		
	Position of the aorta:		
	Position of the PA:		
Ventriculo-arterial connection:	Concordant:		
	Discordant:		
Pulmonary venous return:	Normal:		
	Abnormal:		
	Right veins:		
	Left veins:		
Systemic venous return	Normal:		
	Abnormal:		
	IVC:		
	SVC:		
	LSVC:		
Inter-atrial septum	Bulging:		
	Size of the shunt:		
	Direction of the shunt:		
Interventricular septum	Bulging:		
	Size of the shunt:		
	Direction of the shunt:		
Cardiac chamber			
RA	Dimensions:	Volume	Longitudinal strain
			3D volumes
LA	Dimensions:	Volume	Longitudinal strain
			3D volumes
LV	Dimensions:	M-mode:	Strain: global, septal, lateral
	Wall thickness:	Biplane volumes	3D: volumes, EF, SV
	Systolic function:		
	Diastolic function:		
RV	Dimensions:	2D measures	Strain: global, septal, lateral
	Wall thickness:	Functional indices	3D: volumes, EF, SV
	Systolic function		

Continued

Table 3 Continued

Patient name
Date of birth:
Date of examination:
Age (years/months):
Weight (kg): Length (cm): BSA (m^2):
Arterial pressure (mm Hg): HR: b.p.m.
Oxygen saturation: % Rhythm:
Department: Operator:
Echo machine: Software employed for 3D, strain analysis:

	Basic anatomical/functional detail	Basic measures/functional parameters	Advanced measures/functional parameters
Valves			
TV	Anatomy: Regurgitation: none, trivial, mild, moderate, severe Stenosis: none, mild, moderate, severe	RV–RA pressure difference Annulus diameter Regurgitant parameter Stenotic parameters Power Doppler data	Tissue Doppler data Regurgitant parameters
MV	Anatomy: Regurgitation: none, mild, moderate, severe Stenosis: none, trivial, mild, moderate, severe	RV–RA pressure difference Annulus diameter Regurgitant parameter Stenotic parameters Power Doppler data	Tissue Doppler data Pulmonary vein assessment
Aortic valve	Anatomy: Regurgitation: none, trivial, mild, moderate, severe Stenosis: none, mild, moderate, severe Aortic root and ascending aorta:	Max velocity, max and mean gradient Diameters: annulus, root, junction, Asc Ao, Sub-Ao Regurgitation parameters: Stenosis parameters:	
Pulmonary valve	Anatomy: Regurgitation: none, trivial, mild, moderate, severe Stenosis: none, mild, moderate, severe	Max velocity, max and mean gradient Annulus diameter Regurgitation parameters: Stenosis parameters:	
Great vessels			
Main pulmonary artery		Max velocity, max and mean gradient Diameter	
Right pulmonary artery:		Max velocity, max and mean gradient Diameter	
Left pulmonary artery:		Max velocity, max and mean gradient Diameter	
Aortic arch	Sidedness	Functional parameters Max velocity, max and mean gradient Run-off: Retrograde flow: Diameters at different points	
Arterial duct/collaterals			
Coronary arteries	Origin:	Diameters	
Pericardium/pleura	Effusion/others	Systolic and diastolic diameters	
Abdominal aorta	Flow pattern: normal, demodulated, retrograde, vasoconstriction pattern[a]	Max velocity, acceleration, and deceleration time	
Inferior vena cava/hepatic veins	Excursion Congestion: Reversal flow:	Systolic and diastolic diameters	

Continued

Table 3 Continued

Patient name **Date of birth:** **Date of examination:** **Age (years/months):** **Weight (kg): Length (cm): BSA (m^2):** **Arterial pressure (mm Hg): HR: b.p.m.** **Oxygen saturation: % Rhythm:** **Department: Operator:** **Echo machine: Software employed for 3D, strain analysis:**			
	Basic anatomical/functional detail	**Basic measures/functional parameters**	**Advanced measures/functional parameters**
Others			
Quality of the examination	Acoustic window: poor, sufficient, good, excellent Patient's collaboration: poor, sufficient, good, excellent Completeness of the examination: partial, sufficient, good, excellent		
Conclusions			
Signature:			
Z-score sources:			

2D, two-dimensional; 3D, three-dimensional; IVC, inferior vena cava; EF, ejection fraction; LSV, left superior vena cava; SVC, superior vena cava.
[a]Rapid, steep acceleration and deceleration.

Completeness and conciseness

A report should be complete (including all relevant information) and, ideally, concise (fast to read). For each CHD, it is important to outline the essential anatomical details, measures, and functional parameters to be reported. Supplementary data online, *Table S15*, has been provided a checklist of all essential data to be included in the reporting of major CHDs.

When, what, and how to quantify?

Studies have shown the benefit of quantitative over qualitative evaluation of cardiac defects[4–6] given the significant inter- and intra-observer variability of qualitative assessments, which may lead to misleading interpretation of results.[4–6] What and when to quantify, however, has not yet been completely defined. Latest updates of the Intersocietal Accreditation Commission[5] suggest that numerical data for paediatric transthoracic echocardiograms should include (but not be limited to) measurements of left ventricle (LV) diameters or volumes, LV wall thickness, ejection fraction (EF), and aortic root dimensions.[5] A quantitative measurement of the LV has been also advised during the performance of a targeted echocardiography in the neonatal intensive care unit.[8,12] Whether a basic quantitative evaluation of some cardiac structures should be applied to all subjects (including screening outpatient visit), or only to selected cases, is unclear. Furthermore, no recommendations/consensus of which indices should be evaluated for specific cardiac defects exist.

Measurements of specific cardiovascular indices may raise several practical issues,[6–8,12] especially when dealing with neonates and infants. The level of sedation/cooperation, for instance, is important when performing echocardiographic measurements in children.[6–8,12] When a complete examination is advised (e.g. pre-operative, clinical instability, etc.), one may have to adopt a lower threshold for sedating patients.[6,7] Conversely, sedation may not be a good use of time and resources in case of a screening echocardiography.[7–9]

Clinical Advice:

Strength of advice	A basic screening normal echocardiographic report in neonates, infants and pre-adolescent children may limit to a complete segmental anatomical and functional (systolic and diastolic function, flow across the valve and the main vessels) description without quantification.	
	In older children and/or with the suspicion of congenital or acquired heart diseases a minimum quantitative evaluation is advised over qualitative evaluation.	
	A basic quantification is advised. This should include the evaluation of LV size (diameters, wall thickness, and volumes), LV EF, left atrial (LA) size (area, volume), and aortic diameters. For right ventricle (RV), quantitative evaluation of end-diastolic and end-systolic areas and lengths, basal and mid-diameters, and right ventricle outflow tract diameter, are advised. RV function should be assessed by using fractional area change (FAC), and tricuspid annular plane systolic excursion (TAPSE). Right ventricle to right atrium pressure difference and flow velocities across the aortic valve, pulmonary valve, and aortic arch should also be reported. Measurements should refer to specific age and body size reference values.	
	In uncooperative children, sedation is advised when a complete examination is required (e.g., pre-operative, clinical instability, etc.)	

Gap in knowledge:

The level of sedation/cooperation may alter the quality of examination, and of measurements, and it's difficult to establish when an examination may be of sufficient quality.

Table 4 Major parameters for quantitation in paediatric echocardiography

	Basic measures/functional parameters	Advanced measures/ functional parameters
Cardiac chambers		
RA	AP diameter (mm; Z-score), LL diameter (mm; Z-score), area (cm^2; Z-score):	LS
	Volume: (mL, mL/m^2), EF%	3D
LA	AP diameter (mm; Z-score), LL diameter (mm; Z-score), area (cm^2; Z-score):	LS
	Volume: (mL, mL/m^2), EF%	3D
LV	M-mode:	Strain:
	LVIDd (mm; Z-score), LVIDs (mm; Z-score), IVSd (mm; Z-score), LVPWd (mm; Z-score):	GLS (LS) %
	EF%: Mass (g)	3D:
	Biplane volumes, area, and length:	LVEDV (mL, mL/m^2; Z-score):
	LVEDV (mL, mL/m^2; Z-score):	LVESV (mL, mL/m^2; Z-score):
	LVESV (mL, mL/m^2; Z-score):	EF%, SV (mL, mL/m^2)
	LVEDA (cm^2; Z-score):	
	LVESA (cm^2; Z-score):	
	LVEDL (mm; Z-score):	
	LVESL (mm; Z-score):	
	EF%	
RV	2D measures	Strain:
	RVED area (cm^2; Z-score):	GLS (LS) %, septal LS, lateral LS
	RVES area (cm^2; Z-score):	3D:
	RVED length (mm; Z-score):	LVEDV (mL, mL/m2; Z-score):
	RVES length (mm; Z-score):	LVESV (mL, mL/m^2; Z-score):
	FAC%	EF%, SV (mL, mL/m^2)
	RV1 (mm; Z-score), RV2 (mm; Z-score):	
	Functional indices	
	TAPSE (mm), TDI lateral s′ (cm/s)	
Valves		
TV	RV–RA difference of pressure (mm Hg):	Tissue Doppler (lateral annulus)
	Annulus (mm; Z-score):	e′ (cm/s), a′ (cm/s), s′ (cm/s), E/e′
	Regurgitant parameter:	Regurgitant parameters
	VC (mm), PISA radius, PHT (ms)	EROA (cm^2), 3D VC or EROA (cm^2)
	Stenotic parameters:	
	PHT (ms), valve area (cm^2), EOA (cm^2)	
	Max/mean grad (mm Hg):	
	Inflow velocity time integral (ms), valve area (cm^2)	
	Power Doppler:	
	E (cm/s), A (cm/s), DT (ms), IVRT (ms)	
MV	Annulus (mm; Z-score):	Tissue Doppler
	Regurgitant parameters:	IVS:
	VC (mm), jet area (cm^2), jet length (mm), jet density, PHT (ms)	e′ (cm/s), a′ (cm/s), s′ (cm/s), E/e′
	Stenosis parameters:	Lateral annulus:
	PHT (ms), MVA (cm^2), EOA (cm^2)	e′ (cm/s), a′ (cm/s), s′ (cm/s), E/e′
	Max/mean grad (mm Hg):	Pulmonary vein assessment
	PW Doppler	Ar velocity (cm/s), A duration (ms),
	E (cm/s), A (cm/s), DT (ms), IVRT (ms)	D (cm/s), S (cm/s)
Aortic valve	V max (m/s)	
	Max/mean grad (mm Hg):	
	Annulus (mm; Z-score):	
	Root (mm; Z-score):	
	Junction (mm; Z-score):	
	Asc Ao (mm; Z-score):	
	Sub-Ao diameter (mm; Z-score):	

Continued

Table 4 Continued

	Basic measures/functional parameters	Advanced measures/ functional parameters
	Regurgitation parameters: VC (mm), PISA radius (mm), PHT (ms), EROA (cm^2), reg vol (mL), reg %, jet diameter/LVOT, jet width/LVOT, retrograde flow in Dao, LVEDV Z-score, LVEDV/BSA (mL/m^2) Stenosis parameters: PHT (ms), valve area ($cm^{2)}$, AVA (cm^2), EOA (cm^2) Max/mean grad (mm Hg):	
Pulmonary valve	Annulus (mm; Z-score): V max (m/s) Max/mean gradient (mm Hg): Regurgitation parameters: VC (mm), PHT (ms), jet/annulus width ratio, reversal flow in pulmonary arteries, termination of flow in mid–late diastole Stenosis parameters: PHT (ms) Max/mean grad (mm Hg):	
Great vessels		
Main pulmonary artery	Diameter (mm; Z-score): V max (m/s), peak/mean grad (mm Hg)	
Right pulmonary artery	Diameter (mm; Z-score): V max (m/s), peak/mean grad (mm Hg)	
Left pulmonary artery	Diameter (mm; Z-score): V max (m/s), peak/mean grad (mm Hg)	
Aortic arch	Functional parameters V max (m/s), peak/mean grad (mm Hg) Run-off: Reverse flow: Diameters IA-LCA (mm; Z-score): LCA-LSA (mm; Z-score): After LSA (mm; Z-score): Isthmus (mm; Z-score): Desc Ao (mm; Z-score): Abd Ao (mm; Z-score):	
Arterial duct/ collaterals		
Coronary arteries	LCA (mm; Z-score): LDA (mm; Z-score): Cx (mm; Z-score): RCA (mm; Z-score):	
Pericardium/pleura	Max systolic diameter (mm) Max diastolic diameter (mm)	
Abdominal aorta	V max (m/s), Dec time (ms), Acc time (ms)	
Inferior vena cava/ hepatic veins	Systolic diameter (mm), diastolic diameters (mm)	

Ao, aorta; Abd Ao, abdominal aorta; Ar, peak retrograde flow velocity during atrial contraction; AP, anteroposterior; BSA, body surface area; b.p.m., beats per minute; cm/s, centimetre/second; Cx, circumflex coronary artery; D, peak antegrade flow velocity during ventricular diastole; Desc Ao, descending aorta; DT, deceleration time; EF, ejection fraction; EOA, effective orifice area; GLS, global longitudinal strain; IA, innominate artery; IVC, inferior vena cava; IVS, interventricular septum; IVRT, isovolumetric relaxation time; IVSd, interventricular septum diastolic thickness; LCA, left common coronary artery; LDA, left descending coronary artery; LL, latero-lateral; LS, longitudinal strain; LSVC, left superior vena cava; LVED, left ventricular end-diastolic; LVES, left ventricular end systolic; LS, longitudinal strain; LVPWd, left ventricle posterior wall diastolic thickness; mm, millimetres; PHT, pressure half-time; PISA, proximal isovelocity surface area; RCA, right coronary artery; S, peak antegrade flow velocity during ventricular systole; SCV, superior vena cava; FAC, fractional area change from diastole to systole; TAPSE, tricuspid annular plane systolic excursion; PWD, power Doppler; TDI, tissue Doppler; TV s′, velocity of tricuspid annular systolic motion early diastolic velocity (e′); A′, late diastolic velocity.

Table 5 Example of basic post-surgical report

Patient name
Age (years/months): Weight (kg): Length (cm): BSA (method):
Arterial pressure (mm Hg): HR: b.p.m. Oxygen saturation: Rhythm:
Department: Operator:
Type of surgery: Surgery date:
Echo machine: Software employed for 3D, strain analysis:

Diagnosis

	Anatomy	**Measures/functional parameters**
Pericardium:	Effusion:	Effusion size max systolic (mm)
	Thrombi:	Effusion size max diastolic (mm)
	Inflammation:	
Abdominal aorta pulsatility:	Normal	Ascending/descending times
	Decreased	Reversal flow
Vena cava and hepatic veins	Dimension	Pulsatility
		Reversal flow
Systemic venous return	Obstruction	
Inter-atrial septum	Residual shunt	Shunt size
	Shunt direction	
	Septal bulging	
Interventricular septum	Residual shunt	Shunt size
	Shunt direction	Pressure difference across the defect
	Septal bulging	
Cardiac chambers		
RA	Volumes	
LA	Volumes	
LV	Dimensions:	M-mode:
	Wall thickness:	Biplane volumes
	Systolic function:	
	Diastolic function:	
RV	Dimensions:	TAPSE (mm); TDI lateral s′ (cm/s)
	Wall thickness:	FAC%
	Systolic function	
Valves		
TV	Anatomy:	RV–RA difference of pressure (mm Hg):
	Regurgitation: none, trivial, mild, moderate, severe	Max and mean grad (mm Hg):
	Stenosis: none, mild, moderate, severe	Vena contracta (mm)
MV	Anatomy:	Mitral inflow PW Doppler and tissue Doppler diastolic parameters
	Regurgitation: none, trivial, mild, moderate, severe	Max and mean grad (mm Hg):
	Stenosis: none, mild, moderate, severe	Vena contracta (mm)
Aortic valve	Anatomy:	Max velocity (m/s), max/mean grad (mm Hg):
	Regurgitation: none, trivial, mild, moderate, severe	Vena contracta (mm)
	Stenosis: none, mild, moderate, severe	PHT (ms)
Pulmonary valve	Anatomy:	Max velocity (m/s), max and mean grad (mm Hg):
	Regurgitation: none, trivial, mild, moderate, severe	Vena contracta (mm)
	Stenosis: none, trivial, mild, moderate, severe	PHT (ms)
Great vessels		
Main pulmonary artery	Supravalvular stenosis:	Max velocity (m/s), max and mean grad (mm Hg):
	Stenosis:	Narrowest point (mm)
Right pulmonary artery:	Stenosis:	Narrowest point (mm)
		Reverse flow:

Continued

Table 5 Continued

Patient name
Age (years/months): Weight (kg): Length (cm): BSA (method):
Arterial pressure (mm Hg): HR: b.p.m. Oxygen saturation: Rhythm:
Department: Operator:
Type of surgery: Surgery date:
Echo machine: Software employed for 3D, strain analysis:

Diagnosis	**Anatomy**	**Measures/functional parameters**
Left pulmonary artery:	Stenosis:	Narrowest point (mm)
		Reverse flow:
Aortic arch	Stenosis:	Reverse flow:
		Narrowest point (mm)
Arterial duct/collaterals	Presence	Max velocity (m/s), max and mean grad (mm Hg):
	Origin	Reverse flow:
	Direction	Narrowest point (mm)
Pleural effusion	Present	Maximal diameter
	Absent	
Diaphragmatic movements	Normal	M-mode of diaphragmatic excursions
	Decrease	
	Absent	
	Paradox	
Others		
Quality of the examination		
Conclusions		
Signature:		

BSA, body surface area; b.p.m., beats per minute; FAC, fractional area change from diastole to systole; grad, gradient; PHT, pressure half-time; V max, max velocity; TAPSE, tricuspid annular plane systolic excursion; PWD, power Doppler; TDI, tissue Doppler; RV, right ventricle; RA, right atrium.

Reproducibility and practice

A report format that can be used in various settings (from the intensive care unit to the outpatient department) should be employed.[2,4,7,8,12] A report should be practical, easy to apply, and comprehensible for operators and readers with different levels of experience and skill.[2,4,7,8,12]

Clinical Advice:

Strength of advice	The final report should be easy to understand for professionals of different settings, skills, and experience. In more complex reports, a final short summary may be helpful by providing an overview of the report.	

Ability to evolve over time

New techniques including STE, 3D,[29,31,32] and blood STE[97–108] are gaining consensus, especially in complex cardiac cases.[97–106] Thus, a reporting format should be able to evolve over time, with the inclusion of new parameters, new terminologies, and evolving definitions.[29,31,32,106,107] Given the inter-vendor variability[30] that may generate different ranges of normality, the inclusion of echocardiographic equipment and software employed for complex strain[106,107] and 3D analysis[29,31,32] needs to be incorporated into the report.

Clinical Advice:

Strength of advice	An effective system for reporting in paediatric echocardiography requires flexibility and the possibility to add elements deriving from new or emerging technologies.	

Digital and compatible

Digital era: image analysis and reporting

During the last decade, there has been a progressive transition from analogue to digital echocardiographic laboratories.[3,14,137–141] Digital reporting is superior to traditional videotape and phone-based methods as recently underscored.[3,14,137–141] There are several advantages to a digital system including review, comparison, storage, post-processing, sharing of studies (including in real time through telemedicine), quantitative analysis, and superior resolution.[3,137–141] Furthermore, the creation of an automated report of all the measurements may be easily accomplished, avoiding time-consuming manual transcription.[3,137–141] Automated reports may provide the Z-score for each measurement and allow for comparison with previous examinations (e.g. with superimposed previous values). However, there are downsides to a digital system including the lack of accepted standards and legal, licensure, and billing issues.[137–141] It's important furthermore ensuring compatibility of all the echocardiographic machines with the network and a data management and storage

Table 6 Key elements of a paediatric echocardiographic report

	Anatomical details	Quantitative analysis
Position of the heart and situs	Should always be reported	
AV and VA connection	Should always be reported	
Systemic and pulmonary venous return	Should always be reported	
	The presence of a defect should always be reported	The direction of the shunt and size of defect should be described LV–RV pressure difference should always be described in the presence of a VSD
Atria	Anatomical details should always be reported	Quantitation is advised in case of AV defect or significant shunt lesion
Ventricles	Description of systolic and diastolic function and dimension of LV and RV should always be performed	Quantitation of ventricular size is mandatory in shunt lesions, overload of different nature, valvular lesions, or complex CHD with borderline ventricle Quantitation of ventricular systolic and diastolic function is mandatory when a ventricular dysfunction is suspected clinically or detached during echocardiography or during the follow-up of ischaemic damage of different nature, CM, and myocarditis
AV valves	Anatomical details should always be reported	Quantitation is required in case of stenosis, insufficiency, or left/right disproportion
Aorta and ascending aorta	Anatomical details should always be reported	Quantitation is required in case of stenosis, insufficiency, hypoplasia, or dilatation
Pulmonary arteries	Anatomical details should always be reported	Quantitation is required in case of stenosis/hypoplasia or dilatation
Aortic arch and main vessels	Anatomical details should always be reported	Quantitation is required in case of stenosis or dilatation
Pericardium	Anatomical details should always be reported	Quantitation is required in case of effusion
Abdominal aorta	Anatomical and functional details should always be reported	Quantitation is required in case of systemic hypoperfusion of different nature
Inferior vena cava/hepatic veins	Anatomical and functional details should always be reported	Quantitation is required in case of congestion of various nature

AV, atrioventricular; VA, ventricular–arterial; LV, left ventricle; RV, right ventricle.

system (with sufficient memory, protection, and constant updating). Compatibility of different types of 'DICOM' compression and varying approaches to the processing of Doppler data are other important issues to bear in mind.[14,137–141] There are ongoing efforts to overcome these challenges by scientific societies and industry through the Integrating Healthcare Enterprise (see http://www.cocir.org).[14]

Clinical Advice:

Strength of advice	The building of a digital reporting system is advised and encouraged to save time and avoid manual typing errors.	

Gap in knowledge:

Digital technologies need to evolve in terms of compatibility among different data networks.

Basic elements of a paediatric echocardiographic report

Generalities

Inclusion of demographic data such as age, weight, height, and gender is mandatory[1–12] and of heart rate (HR), blood pressure, oxygen saturation, and respiratory rate is strongly advised.[1–12] Other elements including the examination's medical indication, main diagnosis (if known), and previous interventions[1–12] and ongoing therapy should be reported.

Image quality always needs to be reported.[1–12] Because of the inter-vendor variability of the results, it is important to report the vendor and the software employed for analysis especially when innovative analyses are employed.[29,31,32,97,106,107] The source of nomograms should also be detailed.[18–28]

Key elements of segmental analysis

The key elements following the segmental analysis approach should be reported as shown in *Tables 2–6*.

Conclusions section

In this section, the main diagnosis together with essential functional elements (e.g. the presence and the size of PDA in duct-dependent lesions, the presence and the size of a patent foramen ovale (PFO) in transposition of the great arteries, etc.) should be reported.[1–12] The conclusion should be easy to interpret for all professionals independent of level of seniority, should attempt to answer the pertinent clinical question, and should allow for significant abnormal findings to be clearly communicated.[1–12]

Clinical advice:

Strength of advice	A paediatric echocardiographic report should contain patient's generalities, key elements of the segmental analysis and a concise and easy to understand conclusion section.	

Interpretation of quantitative data

In the following paragraphs, major issues related to the quantification and interpretation of echocardiographic data in the paediatric age are detailed. The projections and the methods for image acquisition and measurement performance have been extensively detailed in previous publications.[1–12] Thus, we'll be limited to discussing issues related to the interpretation of the quantitative data and the choice of the parameters to be used at different ages and in different conditions.

The use of Z-scores

For correct echocardiographic quantification of cardiac structures, it's important to refer to age and body size–specific nomograms.[18–28,47] The choice of nomogram is important, as many earlier nomograms had significant limitations.[18–21] Furthermore, a great variability of results may be observed by using different Z-score sources.[19,21,117]

Major 2D measures

Robust nomograms are currently available for all the major 2D measures (cardiac chamber dimension, area, valvular annulus, aorta, pulmonary arteries, and aortic arch diameters) covering different age, body size ranges, and major ethnicities. When utilizing Z-scores, it is important to know their source and associated limitations.[18,19,21] The use of different nomograms may generate discordant results; thus, multiple sources of Z-scores may be used to have a comparison among them, but during the follow-up, it's important to compare Z-scores from the same source.[18,19,21] Comparing current nomograms has shown the two most recent nomograms (Lopez et al. and Cantinotti et al.)[20,21] have the most comparable ranges of normality with difference limited within a Z-score of 0.5 (Z-score range, 0.001–1.26). Differences were higher at lower extremes of body surface area (BSA), especially for the neonatal age.[20,21] In summary, despite the great advancement in the last years, furthermore, some limitation of Z-scores still exists.[18,19,21] Data are limited for some measures (vena cava, atrial volumes) and some specific sub-groups such as pre-term, low weight birth, and young athletes where the adoption of formulas employed for the whole population may result suboptimal.[18]

Diastolic parameters

For blood flow Doppler and tissue Doppler parameters evaluating diastolic function, actual nomograms present quite reproducible intervals,[18,27,28] except for neonates and infants, where data are limited and contrasting.[18,27,28] Due to the scarce dependence of diastolic values on age and body size, however, normal values are difficult to express as Z-scores, and their expression as mean values plus standard deviation by age groups has been often preferred.[18,27,28]

Newer strain and 3D techniques

Paediatric nomogram on newer STE[18,22,23,26] has been reported both for atria and ventricles. Normal paediatric values on LV and right ventricular (RV) volumes[18,24,25] and LV mass,[18,24,32,47] by 3D echo have also been published, while data on 3D valve size are still limited.[18,31]

Clinical Advice:

Strength of advice	The use of Z-scores is advised for quantification in the pediatric age.	
	Pediatric echocardiographic Z-scores are available for most of the commonly evaluated parameters including newer strain and 3D techniques.	
	Z-scores sources and their strength and limitations should be known. Different Z-score sources may produce different Z-scores for the same measurement in the same patient. The use of most recent normograms is advised, and attention should be paid at BSA extremities where range of normality may diverge in different sources.	
	The same normogram sources should be employed during follow-up for comparison of data.	
	Z-scores obtained by different normograms should be compared, and in case of discordant results the range of Z-scores obtained by different sources should be indicated.	

Gap in knowledge:

- For diastolic parameters, nomograms present limitations because diastolic parameters are less dependent on age and body size.
- Dimensional and functional nomograms are lacking for previously preterm children, low birth weight children, and young athletes.

Ventricular dimensions and function

LV size and systolic function

Quantification of LV dimensions, area, volumes, and function is a basic and fundamental part of the echocardiographic examination at any age.[2,7,10,33] Despite being basic, ventricular measurements in the paediatric age are not completely standardized yet and are subject to a significant inter- and intra-operator variability.[2,7,33] Which method should be employed for LV size and function quantification at different paediatric ages remains a matter of discussion.[2,7,34–36] For years, paediatric guidelines[7] suggested the use of the area–length method for the measurements of LV volumes in the paediatric age, since it's more reproducible.[7,34–36] The biplane Simpson method, however, which is the standard in adults,[33] has been now accepted also for paediatric age.[12,106]

The used methods for LV volume quantification by 2D echocardiography rely on the geometric assumption of a fixed LV shape that, however, may not be applicable in all CHDs.[7,34–36] Furthermore, the LV shape and dimensions are highly variable even at slight angulations of the probe, which may result in indifferent LV diameters and volumes.[7,34–36] As a result, apex foreshortening (both in the apical and subcostal view) and incomplete visualization of endocardial borders are quite common errors in the 2D evaluation of the LV.[7,34–36] Limited corrections for shape distortion are provided by the area–length formula, while the biplane Simpson method allows for a shape correction but still relies only on two planes (four- and two-chamber views).[7,34–36] The use of 3D echocardiography, offering the advantage of not relying on geometric assumptions and being unaffected by apex foreshortening, may provide a better reproducibility and closer agreement with cardiac magnetic resonance imaging (cMRI).[29,37] 3D technology also offers more accurate semi-automated methods for cardiac

chamber dimension, volume, and function that help reduce the intra- and inter-observer variability.[29,37]

RV sizes and systolic function

As for adults,[33] paediatric guidelines[2,7] recommend measuring the RV area, length, diameters (end-diastolic diameters at the basal and mid-cavity levels), and basic functional parameters [FAC (fractional area change from diastole to systole) and TAPSE (tricuspid annular plane systolic excursion)] in an apical four-chamber view. It's well known that RV evaluation by 2D echocardiography suffers from important limitations that are just partly overcome by 3D echocardiography.[2,7,29,33] In fact, RV volumes calculated by 3D echocardiography are not always easy to acquire, due to the poor acoustic window and irregular shape of the RV (especially the RV infundibulum in cono-truncal defects that underwent previous surgery).[2,29,33] 3D echocardiography furthermore underestimates RV volumes compared with cMRI.[7,29,33]

Speckle tracking analysis

The use of STE has gained increased consensus in adults, and the use of global longitudinal strain is currently advised for ventricular function quantification in the adult population and in the adult with CHDs.[33,97,142] STE is gaining popularity also in children with acquired and congenital heart disease for the evaluation of subclinical and regional damage (often not visible with conventional parameters) and to better understand complex ventricular–ventricular interactions.[97,107] STE has been proven its value for early diagnosis of cardiac dysfunction and follow-up in children with cardiomyopathies (CM)[98] and myocarditis.[99] STE, furthermore, may be helpful for follow-up in children who undergone corrective (e.g. tetralogy of Fallot, CoA)[100–102] or palliative (e.g. Fontan circulation)[103,104] surgery as well as in the pre-operative risk assessment.[105] STE is also advised for the LV function assessment and follow-up of children after cardiotoxic chemotherapy.[106]

Clinical Advice:

Strength of advice	For the evaluation of LV volumes and LVEF, the area-length method is advised in neonates and infants, while in older children the Biplane Simpson method is advised.	
	For RV measurements of area, length, diameters, and basic functional parameters (FAC and TAPSE) are advised, with known limitations.	
	The use of LV global longitudinal strain (GLS) and RV GLS should be increasingly advised, but experience and good reproducibility in assessing those parameters is required.	
	In older children and teenagers, 3D echocardiography with LV and RV volume quantification is advised in centers with experience and good reproducibility.	
	3D semi-automated quantification is advised to reduce intra- and intra-observer variability in LV quantitation	

Gap in knowledge:

3D echocardiographic quantification of LV and RV is not advised at this stage in neonates and children.

Diastolic function

Echocardiographic evaluation of diastolic function in children is challenging. Patterns of diastolic dysfunction in children and systems for their classification have not been clearly defined yet.[2,7,18,27,28] Adults' standards for the definition of diastolic dysfunction[56,57] are often employed in the paediatric age, without validation. While their application[56,57] in older children may be acceptable,[7] adult definition is inapplicable in neonates and infants where the pattern of Doppler mitral early diastolic velocity (E)/late diastolic velocity (A) is highly variable, and inversion may be physiological.[18,27,28] At high neonatal HR, furthermore, the phenomenon of E/A fusion is quite frequent.[7,18,27,28]

LV diastolic function

There are limited data comparing echocardiographic parameters to evaluate the diastolic function in children with invasive data and/or clinical outcomes.[47,49–51] Data from children with CM[47,49–51] are limited and contrasting. A large study[51] of 175 children (0–18 years) with different CM showed the inadequacy of adult guidelines for discriminating diastolic dysfunction. Furthermore, quite surprisingly, children with CM had most of the echocardiographic diastolic parameters [isovolumic relaxation time (IVRT), mitral E/A wave Doppler velocities, and e′ tissue Doppler (TDI) velocities] within the paediatric ranges of normality.[51] Left atrial (LA) volume[47,49] and E/mitral annular TDI early diastolic velocity (e′)[49] were higher than in the control group[47,49] in studies on children with different CM. Interestingly, LA strain peak systolic values and LA strain rate were both decreased[49] and able to discriminate between CM and control groups ($P < 0.001$).[49] Evaluation of LA strain also increased sensitivity in the detection of high LA pressure in pre-term infants.[143]

RV diastolic function

Literature about RV diastolic function mainly derives from studies on pulmonary hypertension (PH).[59–63] Similarly, in children with idiopathic PH[63] or mixed[60,61] PH, tricuspid valve (TV) TDI annular e′ tissue Doppler velocities were lower than in controls and correlated with invasive RV end-diastolic and mean pulmonary arterial pressure.[61,63] However, the tricuspid E/e′ ratio did not correlate with RV end-diastolic pressure.[63] RA strain measurements are also useful indicators of RV diastolic function. In adults and in children with PH, all phases of atrial function (reservoir, conduit, and pump phase) have been shown to be impaired.[90] In children and adolescent (2 months–18.0 years) with PH, indices combining data of systolic and diastolic performance such as TV regurgitation (TVR) to TAPSE ratio have shown to correlate with invasive pulmonary vascular resistance index and New York Heart Association class.[62]

Indirect signs of altered diastolic function and co-existing LV and RV diastolic dysfunction

Due to the lack of precise standards, evaluation of LV and RV diastolic dysfunction by indirect parameters may be of great value.[2,7,49–51] LA dilatation, rightward septal bulging, and flow turbulence across the PFO may indicate LV diastolic dysfunction[2,7,49–51] (*Figure 1*). For the RV, common indicators of diastolic dysfunction include RA dilatation, leftward septal bulging, a right-to-left shunt across the PFO, vena cava and hepatic vein congestion, and the presence of pulmonary effusion and ascites[2,7,49–51] (*Figure 2*). Signs of increased pulmonary arterial pressure (increased TV velocity, interventricular septal bulging, right-to-left shunts) and lung congestion are common for both right and left diastolic dysfunctions; therefore, a correct differential diagnosis is important since they may often co-exist.[2,7,49–51]

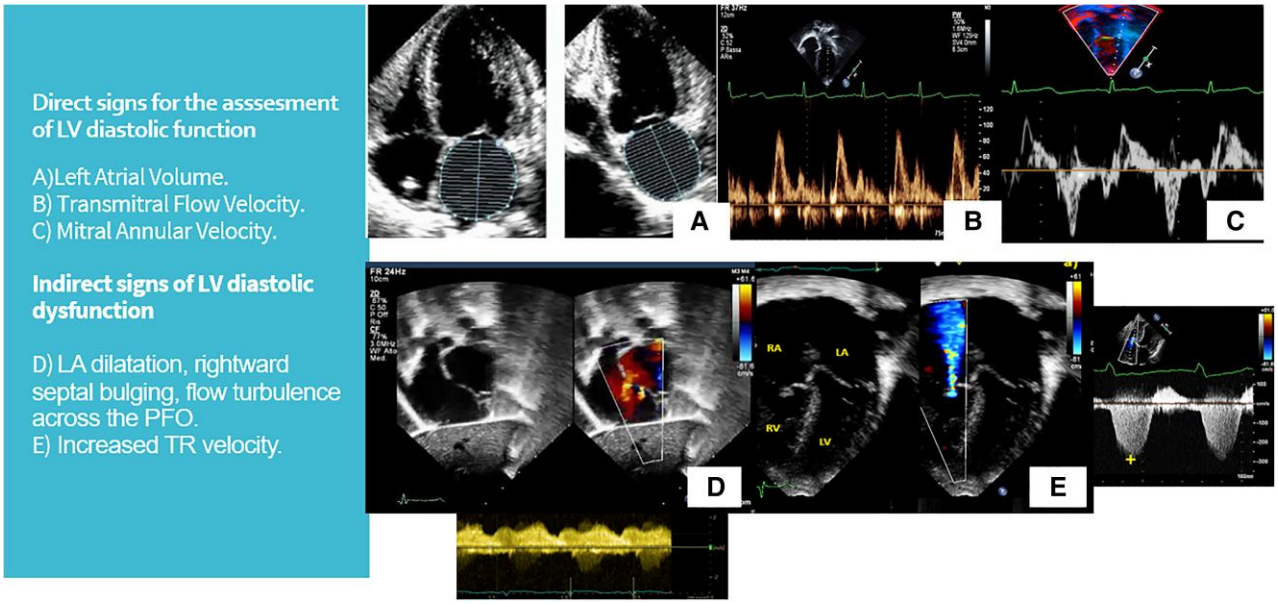


Figure 1 Direct signs for the assessment of LV diastolic function and indirect signs for the assessment of LV diastolic dysfunction. (*A*) Biplane LA volumes. (*B*) Power Doppler transmitral flow velocity. (*C*) Tissue Doppler mitral annulus velocity. (*D*) Subcostal view: LA dilatation, rightward septal bulging, and flow turbulence across the PFO. (*E*) Increased TR velocity. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

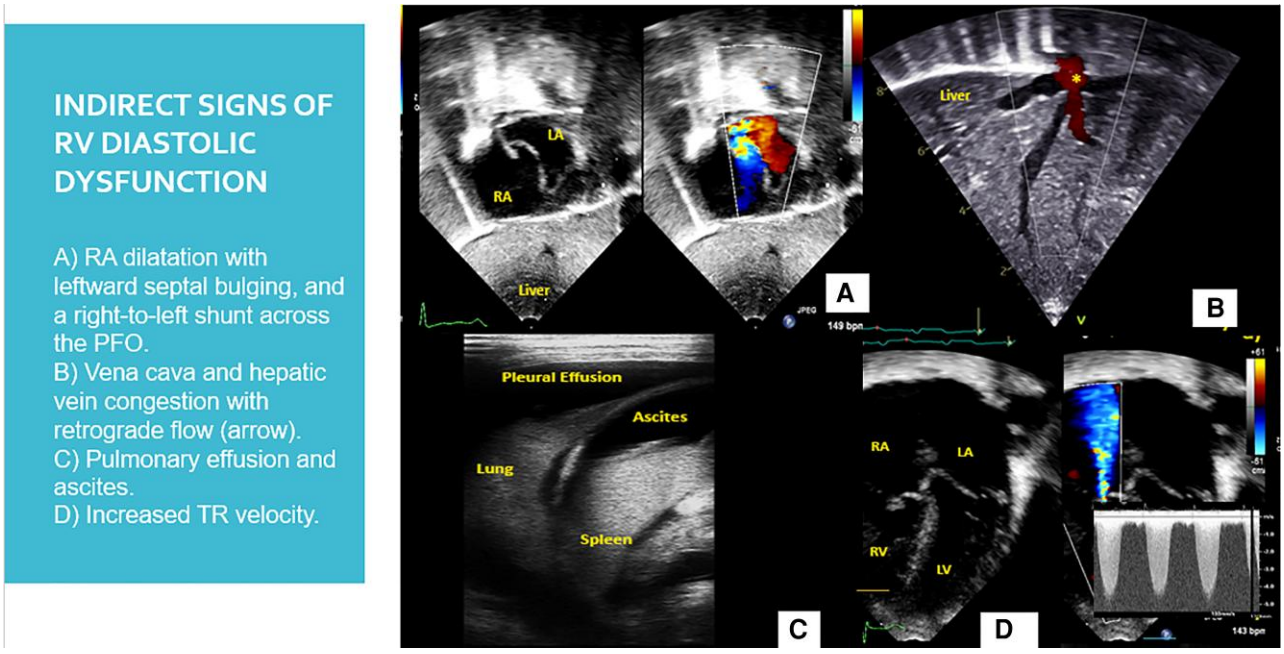


Figure 2 Indirect signs of RV diastolic dysfunction. (*A*) RA dilatation with leftward septal bulging and a right-to-left shunt across the PFO. (*B*) Subcostal view: vena cava and hepatic vein congestion with retrograde flow (arrow). (*C*) Subcostal view: pulmonary effusion and ascites. (*D*) Four-chamber view: increased TR velocity. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Clinical Advice:

Strength of advice	
	Diastolic dysfunction has been poorly defined in neonates and infants. In older children, some of the adult standard parameters may be advised, but they still have significant limitations.
	Evaluation of indirect signs (LA dilatation, rightward septal bulging, flow turbulence across the PFO) are advised for the diagnosis of LV diastolic dysfunction.
	Evaluation of indirect signs (RA dilatation, leftward septal bulging, a right-to-left shunt across the PFO, vena cava and hepatic vein congestion, and the presence of pulmonary effusion and ascites) are advised for the diagnosis of RV diastolic dysfunction.
	Evaluation of signs of increased pulmonary pressure (increase TV velocity, interventricular septal bulging, right-to-left shunts, and lung congestion) are advised for both, the diagnosis of left and right ventricular diastolic dysfunction, which may often coexist.
	Decrease of Doppler/TDI velocities and increase of E/E' ratio compared to normal pediatric value are often, but not universally, encountered in children with diastolic dysfunction.
	TVR/TAPSE is advised for stratification of PH.

Gap in knowledge:

- A system to define and classify diastolic dysfunction in the paediatric age is lacking.
- Despite being promising, the use of atrial strain parameters for the evaluation of the diastolic function requires further validation.

Functional parameters to estimate the severity of valvular lesions

Besides anatomical analysis, a quantitative and semi-quantitative evaluation of disease severity (e.g. the grade of stenosis and/or regurgitation) is also required.[7,12,64] However, an accurate quantification of valve defect severity[7,12,64] remains challenging. Quantitative analysis of AV valve regurgitation in children may be affected by a series of factors, including the difficulty to use fixed cut-off values for a broad range of ages and BSA, the physiological variation of HR and myocardial function with growth that strongly affects Doppler values, the diversity of morphology even within the same defect, and the impact of associated anomalies such as intracardiac shunts.[7]

Semilunar valve: stenotic lesions

For stenotic lesions, only Doppler gradients showed sufficient consistency with invasive gradients [especially for pulmonary stenosis (PS)], while other quantitative parameters used in adults[144] showed significant limitations when applied to children.[65–67,80–82] Doppler gradients, however, are not an exact representation of invasive gradients and require to be interpreted.[7,65–67] Maximal Doppler gradients significantly overestimate, while mean Doppler gradients slightly underestimate invasive peak-to-peak gradients [-6.34 ± 11.9 mm Hg for aortic stenosis (AS) and -6.1 ± 9.4 mm Hg for PS].[4,6] Overestimation of peak Doppler gradients may be partly attributed to the phenomenon of pressure recovery.[65–67,80–82] Pressure recovery is more pronounced in tubular stenosis, as in coarctation of the aorta, while it should be less pronounced in AS, where it should be attenuated by the post-stenotic dilation of the vessel distal to the stenosis, in the ascending aorta.[7,65–67,80–82] Post-stenotic dilatation, however, is less pronounced in children than in adults. Higher paediatric HR, furthermore, may generate higher flow rate, thus enhancing the phenomenon of pressure recovery.[7,65–67,80–82] Thus, correction for pressure recovery is advised when maximal Doppler gradients are used.[7,65–67,80–82] Maximal Doppler gradients corrected for pressure recovery showed limited differences in comparison with mean Doppler gradients (e.g. 1–2 mm Hg both for AS and PS).[7,65–67,80–82] Another important aspect to be evaluated is whether Doppler and invasive gradients are measured in different haemodynamic conditions (awake patient vs. general anaesthesia) since they are influenced by loading conditions and HR and blood pressure may be different in these two settings.[7,65–67] For AS, it is also important to consider the view where the gradient is acquired. Suprasternal view gradients tend to be higher than those acquired in parasternal views; thus, a mean of the two is advised.[7,64]

Heterogeneities remain in the range of Doppler measurements utilized to define mild, moderate, and severe AS and PS.[65–67,69–73,80–82] Few studies have proposed the classification of AS severity in children based on Doppler gradients[72,74,77,79,91] (see Supplementary data online, *Table S16A*). Similarly, there is marked heterogeneity in the classification of PS severity using Doppler gradients, especially in the mild-to-moderate forms.[67] Peak Doppler values ranging from 25 to 40 mm Hg[67–71] have been used to define mild PS, while a mean gradient higher than 50 mm Hg is generally used to define severe PS.[67–71] In addition, Doppler gradients are flow related; thus, in the presence of coexisting lesions such as VSD or MV stenosis, the gradient across the aorta is underestimated.[7] Also, contractility may affect the gradient; thus, in the presence of a reduced SV, the gradient may be underestimated.[7] In this condition, a morphological evaluation of the valve and the complex interplay of the different lesions is advised.[7]

Clinical Advice:

Strength of advice	
	In both AS and PS, indirect assessment of valvular stenosis by evaluation of ventricular response (size and function change over time) and valve morphology is advised.
	The use of mean Doppler gradients is advised since they offer a more precise assessment of invasive gradients despite a slight underestimation.
	For AS and PS, maximal Doppler gradients should be used with caution since they may significantly overestimate invasive peak-to-peak gradient.
	For AS the mean of the gradient obtained in the parasternal and suprasternal view is advised.
	The gradient interpretation should be made with caution since it may be dependent on a series of factors including pre- and afterload, ventricular function, associated shunt lesions, multiple level obstruction, small cavities, anemia, heart rate, and level of sedation/cooperation.
	When peak doppler gradient is used in AS, correction for pressure recovery is advised.

Gap in knowledge:

Classifications of AS and PS severity at different ages are still lacking.

Semilunar valve: regurgitant lesions

In aortic regurgitation (AR), systems to classify disease severity are limited,[50–53] and there is weak evidence supporting the use of quantitative or semi-quantitative parameters commonly employed in adults,[7,144] even after correction for BSA.[7,76–79,81] A few studies tried to compare

some of the most used echocardiographic parameters with those from cMRI[76–79,81] (which is the gold standard) or catheter angiography[82] (see Supplementary data online, *Table S16B*). In a large study with over 135 patients with various CHDs before and after repair/palliation,[77] it has been shown that aortic regurgitant fraction, parasternal vena contracta indexed by BSA, and the ratio of thoracic and abdominal aorta antegrade to retrograde flow and the jet cross-sectional area correlated with the regurgitant fraction measured by cMRI.[77] The ratio of aortic antegrade/retrograde was used also in other smaller studies showing good correlations with the regurgitant fraction derived from cMRI.[76,77] There is an agreement that assessment of LV dilatation by LV Z-scores is of paramount importance for estimation of AR impact on cardiac function and its tolerance over time.[77,78] The presence of pandiastolic reverse flow in abdominal aorta and in descending aorta are generally considered markers of severe AR. Increased cardiac output (to maintain an adequate flow) is another marker of severe AR.[77,78]

Various echocardiographic semi-quantitative and quantitative indices for pulmonary regurgitation (PR) have been evaluated in patients with repaired tetralogy of Fallot[83–86] and compared with cMRI (which represents the gold standard).[85,86] In adults, flow reversal in the main or branch pulmonary arteries, PR jet width of 50% of the pulmonary annular diameter, and PR pressure half-time (PHT) <100 milliseconds (ms) are independent predictors of severity.[84–86] A PR duration of 80 ms and PHT of <100 ms accurately predicted angiographically severe PR in adults.[85,86] Other markers of PR severity have been evaluated both in children[85] and in adults.[86] In children with repaired tetralogy of Fallot, the ratio of diastolic and systolic velocity time integral of main pulmonary artery flow is an index of PR and modestly correlated with RV myocardial performance index EF.[85] Vena contracta has also been used to quantify PR.[85,86] It has been shown that 3D vena contracta correlates well with 2D jet width.[84] However, it's important to remember that 3D colour frame rate is often too slow to properly quantify regurgitation in children.[84]

Clinical Advice:

Strength of advice		
	In both AR and PR, indirect assessment of valvular regurgitation by evaluation of ventricular response (size and function change over time) and valve morphology is advised.	
	Evaluation of PR multiple parameters (jet size and duration, PHT, reversal flow in pulmonary arteries) is advised.	
	For AR, only a few echocardiographic parameters such as vena contracta/BSA, abdominal aorta anterograde to retrograde flow ratio, and a pandiastolic reverse flow in abdominal aorta or descending aorta, have been validated for quantification and may be advised for severity estimation in the pediatric age group.	

Gap in knowledge:

- Echocardiographic recommendations to classify AR severity are lacking.
- Definition of severe PR is clear, while the definition of moderate and mild PR is less well defined.
- Larger studies to evaluate echocardiographic parameters for a more complete and precise assessment of semilunar valve defect in paediatric age are warranted (especially for AR), and a system that classifies severity needs to be developed.

AV valves: stenotic lesion

For stenotic AV valve lesions,[87–89] no clear categorization based on transvalvular echocardiography–derived 'gradients' has been consistently applied to define mild, moderate, or severe obstruction across different paediatric age ranges.[87–89] While various anatomical classifications have been proposed to classify mitral stenosis,[87–89] only a few paediatric studies, however, proposed mitral valve (MV) stenosis classifications according to gradients derived either by pulsed Doppler[87] or cardiac catheterization.[88] The range of gradients proposed to define mild, moderate, and severe mitral stenosis, however, is variable between studies[87,88] and differs from adult recommendations[144] (see Supplementary data online, *Table S16A*).

Quantitative parameters such as valve area[144] have also been poorly validated in children. All quantitative parameters, in fact, are affected by significant physiologic variations with growth, and thus, cut-off values to estimate disease severity (if applicable) should be adjusted for age and body size.[7,87–89] The high HR in children may augment the transvalvular gradient and limit the accuracy or PHT and the effective valve area by the continuity equation.[7] The impact of co-existing shunts [e.g. VSDs that may increase transmitral flow or ASDs that may reduce MV flow by permitting shunting to the right atrium (RA)] also needs to be considered.[7,87–89] Evaluation of atrial size, ventricular size and function, and the presence of ASD is also of importance for AV stenosis evaluation.[7] RV pressure, the presence of hepatic congestion, and the characteristics of the shunt across the PFO are also of relevance.[7]

Clinical Advice:

Strength of advice		
	Indirect assessment of AV stenosis severity by evaluation of the atrial size, right ventricular pressure, RV dilatation and function, hepatic congestion, and the characteristics of the shunt across the PFO is advised.	
	A series of factors that may significantly affect the pressure gradients including the high heart rate, the presence of associated defects that may reduce or increase the preload and/or afterload, the level of sedation/cooperation, need to be carefully evaluated.	
	In the absence of validated pediatric cut-off values, adult criteria for AV valve stenosis estimation may be adopted with caution and awareness of their limitations especially for lower ages (e.g., neonates and children).	

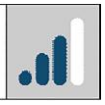
Gap in knowledge:

- There is a lack of validated quantitative/semi-quantitative parameters to classify AV valve disease severity in paediatric patients.
- Studies using both 2D and 3D echocardiographic parameters for the evaluation of the degree of AV regurgitation in comparison with MRI data are advised.

AV valves: regurgitant lesion

For AV regurgitant valve lesions, there are no clear criteria to grade disease severity in the paediatric age group at present.[7,90–96] Quantitative or semi-quantitative indices deriving from adults[144] are commonly employed in children, despite the fact they are not validated and often inapplicable in the paediatric age group.[7,90–96] Vena contracta, one of the easiest and most employed indices used in adults,[144] has been validated only in one study including 149 infants with left AV valve regurgitation after biventricular correction for AV septal defect.[90] The lateral, anteroposterior, and cross-sectional vena contracta area indexed for BSA correlated moderately with Z-scores of LV end-diastolic volumes and showed high inter-observer agreement.[90] Other indices including regurgitant fraction,[94] proximal isovolumetric area,[94,96] and effective regurgitant area by 2D[9,96] and 3D echocardiography[95] have been tested only in studies with small sample sizes.[95,96] As for stenosis, indirect assessment of AV regurgitation severity by evaluation chamber size and RV pressure is important.[7]

Clinical Advice:

Strength of advice	In the lack of validated parameters for direct assessment of AV regurgitation severity estimation in children, indirect evaluation of chamber size and right ventricular pressure is advised 

Gap in knowledge:

- Adult quantitative parameters for AV regurgitation estimation (vena contracta, vena contracta indexed by BSA, regurgitant fraction proximal isovolumetric area, effective regurgitant area by 2D and 3D echocardiography) have been poorly validated in children.
- No clear criteria to grade disease severity in the paediatric age group are available at present.

Estimation of left-to-right shunt severity

Classification of common left-to-right shunt lesions such as septal defects and PDA into mild, moderate, and severe is commonly employed despite the lack of validated criteria.[28,29,39–46] Maximal 2D defect diameter has been used to define small, moderate, and large atrial and septal defects[28, 39–46] (Supplementary data online, *Table S17*), but cut-off values vary among different authors. On the other hand, fixed cut-off values may be inadequate for children, where septal defect size should consider body size and the relation to dimensions of other cardiac structures.[7,28,39–46] The use of 2D measures furthermore may be inaccurate for the assessment of complex geometry of septal defects, where 3D measures may best fit. Criteria for image acquisition and septal defect measurements by 3D echocardiography need to be standardized yet.[29] Echocardiography provides inaccurate estimation of systemic to pulmonary flow ratio (*Qp*/*Qs*), overestimating the degree of left-to-right shunt, compared with cardiac catheterization and cardiac MRI.[7,28] Thus, indirect signs of defect severity including cardiac chamber enlargement, left-to-right chamber dimension ratio, pulmonary artery pressure, and electrocardiographic alterations are of importance for defect severity estimation.[7,28,39–44]

Lastly, the definition of a restrictive PFO should be mentioned, a condition which may require an urgent diagnosis in cyanotic CHD in the neonatal age. This condition has been differently defined based on shunt size and flow velocity[45,46] (see Supplementary data online, *Table S17*). Regardless of the definition used, assessment of direct (turbulent flow) and indirect signs (rightward septal bulging, pulmonary vein dilatation) is essential for the recognition of a restrictive PFO.[7,45,46]

Clinical Advice:

Strength of advice	Indirect evaluation of volume and pressure overload is advised for shunt evaluation, instead of measurements restricted to defect size alone. 
	Assessment of direct (turbulent flow) and indirect signs (rightward septal bulging, pulmonary vein dilatation) is important for the recognition of a restrictive PFO. 

Gap in knowledge:

Systems to define left-to-right shunt severity in relation to body size and to cardiac chamber overload are lacking.

Prediction scores

Risk scores for biventricular repair in complex CHDs

In Supplementary data online, *Table S18*, major risk scores[109–115] in CHDs with borderline LV function of different aetiology including critical AS,[109–115] critical LV outflow tract (LVOT),[109,122] and obstruction at multiple levels[109,122] have been reported (*Figure 3*). Furthermore, we reported parameters for risk estimation of biventricular repair (BVR) in the borderline LV in unbalanced AV septal defect with left dominance (see Supplementary data online, *Table S4*). These include AV valve index, LV inflow index, RV or LV inflow angle, left AV valve reduction index, and VSD size[115–122] (*Figure 4*). Similarly, parameters used to indicate[123–128] the risk for BVR or pulmonary flow augmentation in complex CHDs with borderline RV such as pulmonary atresia with intact ventricular septum and critical PS (see Supplementary data online, *Table S11*) are shown. These include TV Z-score, RV/LV anteroposterior and lateral diameter ratios, RV and RA area, and direction of PFO shunt, as well as tricuspid regurgitation characteristics[123–128] (*Figure 5*).

The use of risk scores for BVR in complex CHDs may be helpful; however, it's important to remark how they all present significant underlying limitations[109,122] that may limit their clinical significance. These include the retrospective design during score development (for all the studies), heterogeneity in echocardiographic parameters evaluated, variability in the definition of outcomes, differences in adopted surgical and interventional strategies, institutional differences, and limited follow-up (e.g. from 1 month to 5 years).[109,122] Most of the scores furthermore were developed in the past two decades and may have limited clinical significance nowadays.[109,122] As a result, their applicability remains questionable.[109,122]

The use of 3D echocardiography may allow a better estimation of LV volumes that are constantly underestimated by 2D echocardiography compared with cMRI.[29,37] The use of 3D echocardiography, furthermore, may also help in a more precise assessment of MV annular dimension, which is often underestimated by 2D measures.[7] Thus, the use of 3D echocardiography may allow BVR in a greater percentage of children as some children are unfairly precluded due to underestimation of LV size by 2D measures.[112]

Clinical Advice:

Strength of advice	Scores for BVR risk prediction may be advised as adjunctive data in complex CHD with diminutive LV, but their clinical utility remains uncertain. 
	3D echocardiography may provide a better assessment of LV volumes and MV dimensions that are often underestimated by 2D measurement. 

Gap in knowledge:

Large and prospective multicentre studies with clear definition of echocardiographic parameters, use of 3D echocardiography, and clear definition of outcomes are required for the development of accurate risk prediction models for BVR.

Risk score for prediction of postnatal CoA in the case of a big arterial duct

De novo diagnosis and/or confirmation of prenatal suspicion of CoA in the presence of a PDA in the first days of life is often challenging. Besides prenatal scores, a series of scores for the prediction of postnatal CoA in the presence of a PDA have been proposed.[129–134] These include (i) the carotid–subclavian artery index (CSAi), (ii) the isthmus/descending aorta diameters (I/D ratio), and (iii) the coarctation probability model (CMP) (see Supplementary data online, *Table S4B*). These scores have been tested in small and relatively small studies enrolling 23–80 neonates. Accuracy of the scores seems to be promising with area under the ROC curve (AUC) varying from 0.96 for CMP to 0.91 for CSAi and up to 0.69 for I/D ratio. Sensibility (87–100% for CSAi, 32.5–91.7% for I/D ratio, 92.7% for CMP) and specificity (69–96% for CSAi, 100–23% for I/D ratio, 94.6% for CMP) were also good.[129–134]

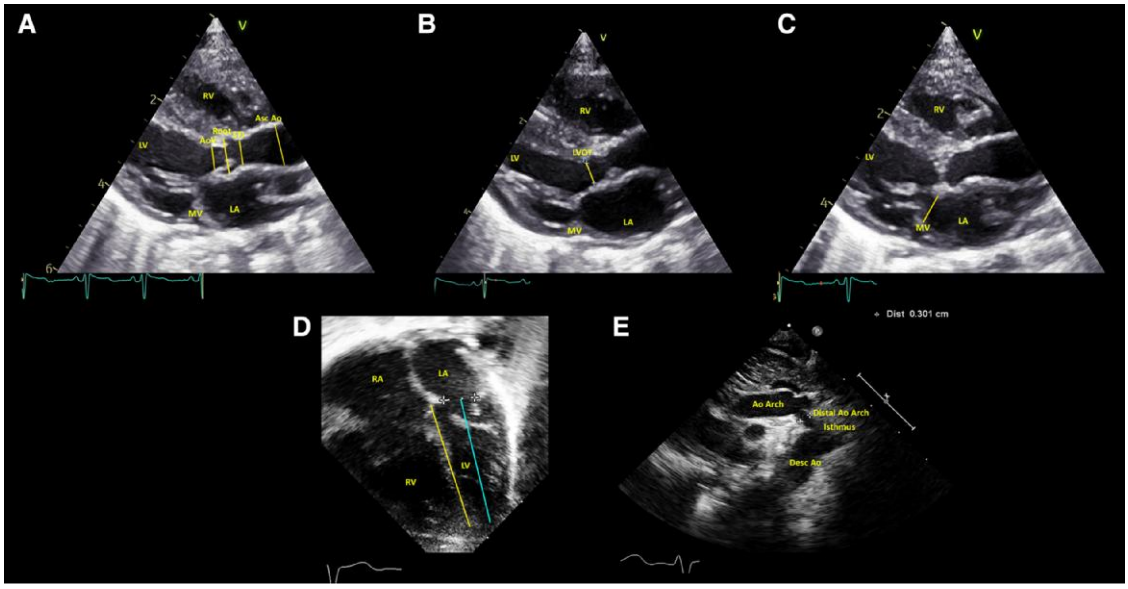


Figure 3 Echocardiographic measures required for risk prediction of borderline LV by the Congenital Heart Surgeons' Society calculator. (*A*) Parasternal long-axis view: an aortic valve annulus, aortic root, sino-tubular junction, and ascending aorta. (*B*) Long-axis view: LVOT. (*C*) Long-axis view: MV annulus. (*D*) Apical four-chamber view: heart long axis (line, from the crux to the apical endocardium) and LV long axis (from the MV plane to the apex). (*E*) Suprasternal view: mid-aortic arch. Ao arch, aortic arch; Desc Ao, descending aorta; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; MV, mitral valve; RA, right atrium; RV, right ventricle, STJ, sino-tubular junction.

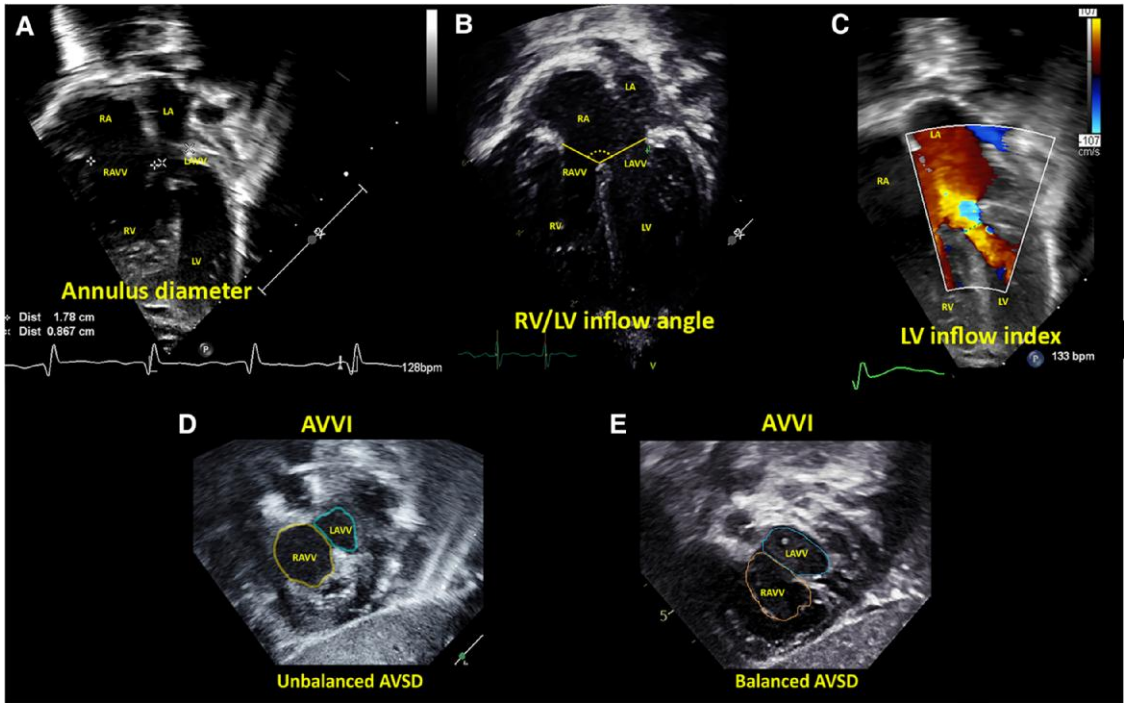


Figure 4 Parameters for the risk prediction of unbalanced AVSD. (*A*) Apical four-chamber view: LAVV and RAVV virtual diameter. (*B*) Apical four-chamber view: the RV/LV inflow angle. (*C*) Apical four-chamber view: the true LAVV annulus evaluated by colour flow (e.g. the LV inflow index). (*D*) Subcostal views: right-to-left AVV valve area ratio of an unbalanced AVSD. (*E*) Subcostal view: right-to-left AVV valve area ratio of a balanced AVSD, respectively. LA, left atrium; LAVV, left AV valve; LV, left ventricle; RA, right atrium; RAVV, right AV valve; RV, right ventricle.

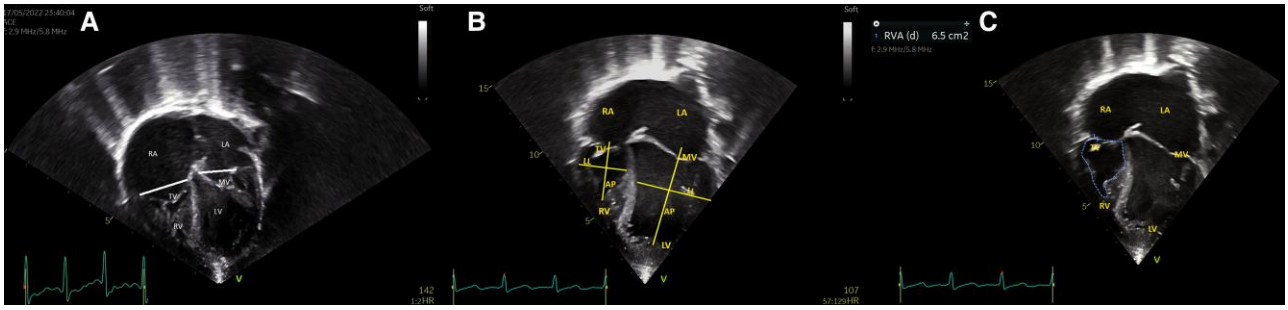


Figure 5 Echocardiographic parameters for risk prediction in borderline RV. A case of pulmonary atresia with diminutive RV. (*A*) TV diameters. (*B*) Base–apex and later lateral diameters of RV and LV. (*C*) RV end-diastolic area. AP, anteroposterior; LA, left atrium; LL, latero-lateral; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle; TV, tricuspid valve.

One of the difficult aspects in the postnatal diagnosis of CoA is to differentiate among the physiological postnatal RV prevalence and a pathological RV to LV disproportion.[129–134] A LV–RV end-diastolic area ratio in a four-chamber view ≥1.3 has been suggested to represent an accurate marker (AUC 0.97) for the need of intervention in an antenatal suspicion of CoA.[129–134] Recent studies outlined the importance of the RV and LV function estimated by STE, as new indicators for the risk of development of CoA.[133,134]

Clinical Advice:

Strength of advice	Risk scores may be advised to increase the accuracy in the diagnosis of aortic postnatal CoA in presence of PDA, together with the evaluation of RV and LV size and function, and direct visualization of posterior shelf.

Gap in knowledge:

Clear criteria for diagnosis of postnatal CoA in the case of a big arterial duct are still lacking.

Conclusion and limitations

The present consensus paper represents a tool that intends to help the clinician in the reporting of normal screening examination and major congenital cardiac defects. Indications for interpretation of echocardiographic measures at different ages and body sizes according to current Z-scores are shown, with a special attention to functional data. Limitations in the evaluation of diastolic function, severity of valvular defects, and shunt lesions in the paediatric age group are highlighted. The examples providing standardized reporting formats intend to improve quality, promote standardization, save time, and assist in teaching and research purposes. These formats may be modified and implemented according to institutional requirements and the availability of new echocardiographic techniques and parameters.

Supplementary data

Supplementary data are available at *European Heart Journal - Cardiovascular Imaging* online.

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Data availability

No new data were generated or analysed in support of this research.

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