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Paediatric Palliative Care in Children with Complex Heart Disease – When to refer?

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PAEDIATRIC PALLIATIVE CARE IN CHILDREN WITH COMPLEX HEART DISEASE – WHEN TO REFER? / CUIDADOS PALIATIVOS PEDIÁTRICOS EM CRIANÇAS COM CARDIOPATIAS COMPLEXAS – QUANDO REFERENCIAR?

Systematic Review

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List of Abbreviations and Acronyms

AHD Advanced heart disease

CAPC Center to Advance Palliative Care

CCC Complex chronic condition

CPP Cuidados paliativos pediátricos

CHD Complex heart disease

CICU Cardiac intensive care unit

ECMO Extracorporeal membrane oxygenation

EOL End of life

HD Heart disease

HF Heart failure

HLHS Hypoplastic left heart syndrome

ICD Implanted cardiac defibrillator

ICU Intensive care unit

MeSH Medical Subject Headings

PPC Paediatric palliative care

PaPaS Paediatric Palliative Screening

PRISMA Preferred Reporting Items for Systematic Reviews and Meta-Analyses

QoL Quality of life

SVHD Single ventricle heart defects

VAD Ventricular assist device

WHO World Health Organization

Wk Week

Wks Weeks

W/O Without

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Abstract

Introduction: Children affected by complex heart disease (CHD) have lifelong risks for morbidities and life-threatening events. Literature is currently scarce and no specific guidelines for paediatric palliative care (PPC) referral exist, whereby, our purpose was to create guidelines for PPC referral of this population in clinical practice.

Methods: A systematic review of the relevant literature published between 2013 and 2023 was performed, in accordance with the protocol of Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA), in PubMed, Embase and Web of Science. The following combination of terms was used: Paediatric Palliative Care referral criteria AND (children with complex heart disease OR complex cardiac disease). Of a total of 43 articles, 11 were considered eligible by the inclusion and exclusion criteria. All significant data was gathered and condensed in a cohesive workflow of PPC and advance care planning.

Results: Of a total of 43 articles, 11 were found to be eligible, based on the inclusion and exclusion criteria defined. Most articles evidenced a consensus regarding the referral of children presenting with single-ventricle cardiac conditions; pulmonary vein stenosis; pulmonary hypertension; symptomatic heart failure; myocardial dysfunction; associated trisomy 13 or 18; mechanical circulatory support; or heart transplant to a specialised PPC team. Treatment and care burden and the use of an implantable defibrillator or other advanced therapies were more controversial and cited in a small number of articles. In dubious cases and concomitant diagnoses or comorbidities, the general criteria for referral to PPC and the Paediatric Palliative Screening Scale (currently validated in portuguese) must be considered.

Discussion and conclusions: Although generalised criteria for referral to PPC have been defined, reflection based on the diagnosis-related group may raise doubts, particularly when invasive curative procedures are available. Despite the scarcity of robust literature, general criteria have been found to exist. In this sense, the authors have established an algorithm for referral to PPC and stressed the importance of timely referral for avoiding unnecessary procedures and ensuring the adoption of a treatment approach suited to the needs of the children and their families.

Future studies are required in order to analyse the applicability of this algorithm in paediatric cardiology settings and to assess the impact of timely referral of children with CHD on quality of life, symptomatic treatment and alignment between care plans and the wishes and choices of patients and their families.

Keywords: Complex heart disease, paediatric palliative care, referral criteria, advance care planning

Resumo

Introdução: Crianças com cardiopatias complexas têm alto risco de comorbilidades e eventos ameaçadores de vida. Atualmente a literatura é escassa e não existem orientações específicas para o seu encaminhamento para cuidados paliativos pediátricos (CPP). Assim, este trabalho teve como objetivo criar normas para a referenciação aos CPP na prática clínica.

Métodos: Foi realizada uma revisão sistemática da literatura relevante publicada entre 2013 e 2023, de acordo com o protocolo do Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA), na PubMed, Embase e Web of Science. A seguinte combinação de termos foi usada: Paediatric Palliative Care referral criteria AND (children with complex heart disease OR complex cardiac disease). Toda a informação significativa foi analisada e condensada num algoritmo coeso de referenciação aos CPP.

Resultados: De um total de 43 artigos, 11 eram elegíveis pelos critérios de inclusão e exclusão. A maioria foi consensual em incluir crianças com uma condição cardíaca envolvendo ventrículo único, estenose da veia pulmonar, hipertensão pulmonar, insuficiência cardíaca sintomática, disfunção miocárdica, trissomia 13 ou 18 associada, sob suporte circulatório mecânico ou transplante cardíaco na referenciação a equipa especializada de CPP. A sobrecarga de tratamento e a utilização de desfibrilador implantável ou outra terapêutica avançada foram critérios menos consensuais por serem referidos numa minoria de artigos. Em casos dúbios ou na presença simultânea de outros diagnósticos ou comorbilidades, devem ser considerados os critérios generalistas de referenciação a CPP e a *Paediatric Palliative Screening Scale* (atualmente validada para português).

Discussão e conclusão: Apesar de haver critérios generalistas de referenciação a CPP, a reflexão tendo em conta o grupo diagnóstico pode suscitar dúvidas, sobretudo em áreas com grande investimento em procedimentos invasivos, com possibilidade de cura. Apesar da escassez de literatura robusta, existem critérios praticamente transversais para referenciação a CPP, tendo as autoras criado um algoritmo de referenciação a CPP, enfatizando a importância da referenciação atempada como ferramenta para uma intervenção mais adequada às necessidades da criança e família, e evitando procedimentos fúteis.

Será importante analisar, em estudos futuros, a aplicabilidade deste algoritmo nos serviços de cardiologia pediátrica, e o impacto da referenciação atempada nas crianças com cardiopatia complexa, nomeadamente na sua qualidade de vida, controlo de sintomas, sobrecarga do tratamento, e adequação dos planos antecipados de cuidados aos seus desejos e escolhas, assim como da sua família.

Palavras-Chave: Cardiopatia complexa, cuidados paliativos pediátricos, critérios de referenciação, planeamento antecipado de cuidados

1. Introduction

Complex chronic condition (CCC) is defined by Feudtner C. *et al* as “any medical condition that can be reasonably expected to last at least 12 months (unless death intervenes) and to involve either several different organ systems or one system severely enough to require specialty paediatric care and probably some period of hospitalization in a tertiary care center”[1].

Around 15% to 20% of children with major heart conditions have high-risk complex cardiac conditions[2]. Prenatal detection of complex heart disease (CHD), defined as lesions requiring surgical intervention or catheterization during the first year of life or requiring multiple staged operations[3-5], but with an important heterogeneity[6], has increased in the past few decades, with heart failure (HF) representing a growing cause of hospital admissions in children[7-9].

Significant medical and surgical advances have resulted in a growing population of children with advanced heart disease (AHD)[8], defined as severe symptoms frequently requiring high technology therapies[10], sometimes intensive, characterized by periods of stability interrupted by acute decompensations that may be mitigated, at least temporarily, by medical and surgical intervention[11,12]. AHD is mainly caused by congenital heart disease (HD), which also accounts for 1/3 of all major congenital abnormalities, and cardiomyopathy (the leading indication for heart transplant during childhood)[13].

Single ventricle heart defects (SVHD) encompass a wide range of cardiac malformations in which there is no way to “fix” or repair the heart back to normal physiology[14], occurring when one of the lower chambers or a valve of the heart is underdeveloped or missing[15]. Hypoplastic left heart syndrome (HLHS) is an example and one of the most severe forms[14]. CHD, particularly SVHD, are stressors, especially for parents[14], which can make decision-making, advance care planning, and communication with families more difficult. There are also intrinsic challenges in prognostication for patients with CHD, not always clear at the time of diagnosis[16].

It is common for a child with CHD to experience more than one life-threatening event, where parents are given a dismal prognosis, yet the child survives. Children benefit from a unique capacity for recovery, growth and developmental progress, even after major neurologic events. However, they have lifelong risks for morbidities and life-threatening events, involving multiple instances of complex decision-making[17]. As a result, children requiring a surgical or trans-catheter intervention before the initial discharge from hospital after birth can be considered to have life-threatening illness[18], which often brings a symptom burden that weighs heavily on children and families[19]. In any case, both children with life-threatening and life-limiting illness present a wide variety of diagnoses and disease courses, with the individual survival varying from hours to more than 20 years[20].

Despite the severity of CHD, it is also important to take other associated comorbidities into account, namely perinatal disorders, birth complex defects, chromosomal abnormalities and polymalformative syndromes, which often involve specific cardiovascular and non-cardiovascular comorbidities that entail significant risks, affecting multiple organ systems[21] and leading to a higher mortality rate[22]. These conditions, particularly trisomy 13 and 18, are the main complex clinical conditions causing death in these children, and can, by themselves, worsen the baseline prognosis and require an individualized advanced care plan[23].

Treatment for children with CHD and developments in medical and surgical technology have increased their survival[3,6,14,24], although at the cost of an increase in disease-related morbidity and significant disability (prolonged hospitalizations with larger number of readmissions[23,25], HF, reoperation, impaired exercise tolerance, multiple painful interventions and technological dependence)[3,14,18,26,27]. In the children affected with CHD whose condition deteriorates quickly and/or unexpectedly, confrontation with death may be far more abrupt[6].

It is believed in Western Society that the primary and only goal of the health system is to achieve cure, causing families and healthcare professionals to experience frustration when faced with incurability and end of life (EOL). This consternation becomes even more significant when the patient in question is a child[23]. The tiring routine of constant visits to the hospital has serious consequences for the child-family binomial, namely anguish due to the lack of clarity in some diagnoses, fear of the possibility of treatment failure, financial maladjustment, distance between the child and the caregiver and stress, among many other damages to the binomial's quality of life (QoL), which are aggravated by harsh hospital routines[23].

From the Latin "*palliare*", meaning "to cloak"[19,28], the World Health Organization (WHO) defines Paediatric Palliative Care (PPC) as an approach that improves the QoL of paediatric patients[26], their families and caregivers in case of life-limiting (in whom premature death is usual, but not necessarily predictable or imminent)[29] and life-threatening (in which there is a high probability of premature death, but also a chance of long-term survival to adulthood)[29] illnesses[29,30]. This approach focuses on the prevention and relief of physical, psychological, social and mental suffering, implementing the holistic and active care concept of treating affect children as a "whole", rather than from an organ system perspective, making them feel more like a person than a patient[3,17,28,31]. PPC delivered simultaneously with disease-directed care[25] has been shown to have significant potential benefits, by managing symptoms, stress and anxiety, improving functional status, reducing the frequency of procedures, and helping with goals of care, informed decision-making, prognostic awareness, advance care planning and communication[3,17,19,26,31-35].

Despite the increasing interest in incorporating PPC over the past years[3,17] and growing research, which currently plays an important role in adding value to healthcare interactions,[19] the issues related to the adequate time for referral remain largely unexplored, which hinders the increase in referral rates[32,36,37]. Up to this date, PPC has been infrequently included in the care of children with CHD; when included, consultations occur mostly too late or not at all[11,38,39].

International standards for PPC were recently revised by Benini F. *et al*[40] and include general conditions (non disease-specific) for eligibility to specialized PPC teams. Thus, PPC should also be provided when any of the following eligibility criteria are met:

1. Hospitalizations: ≥ three for serious clinical crises over a period of six months; > three weeks without clinical improvement; or admission to intensive care unit (ICU) for > one week without clinical progress;
2. A child with difficult and complex management of care handover between the hospital setting and home;
3. Difficulties in making significant decisions and achieving consensus between the child, family and/or medical team on treatment and goals of care;
4. The anticipation of special support during the bereavement period.

Additionally, Paediatric Palliative Screening (PaPaS) Scale[41] is an instrument created by Berstraesser *et al* to facilitate timely and appropriate involvement of PPC to improve the care of children with life-limiting conditions.

The final version of the PaPAs Scale is based on 5 aspects:

1. Course of the disease and impact on the child's daily activities;
2. Expected outcome and burden of treatment;
3. Symptom and problem burden;
4. Preference of health professionals;
5. Estimated life expectancy and the 'surprise question'.

Each aspect was divided into two to five questions for a total of 11 items. Each item includes several options that score from zero to four. Higher individual or total scores should suggest a greater need for PPC. Forwarding to PPC involves a stepwise approach, with three levels of care:

1. Score ≥ 10: considering the introduction of the concept of mortality and PPC, explaining it to the family, creating a plan and discussing how the team is going to include PPC;
2. Score ≥ 15: preparing a PPC approach with basic symptom management alongside treatments to control the disease;
3. Score ≥ 25: PPC becomes the focus of care.

This tool has recently been translated, culturally adapted and validated to the portuguese paediatric population by Palaré, M. J. *et al*[42] and is very beneficial regarding those dubious situations, with particular emphasis on more subjective aspects, such as impact on the daily activities, psychological distress and the preferences/needs of children and families, in addition to the preferences of health professionals, mainly when consensus is difficult.

The diagnosis alone should not represent the only eligibility criteria for PPC, health care needs and prognosis should also be considered[40].

The purpose of this review was to update the main eligibility criteria and recommendations for referral of children with CHD to PPC, based on a critical revision of the available scientific evidence, and to create an objective and practical tool for clinicians.

2. Methods

2.1. Study Characterization

In accordance with Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines, this systematic review's protocol was registered with the International Prospective Register of Systematic Reviews on 12 February 2023, and was last updated on 27 February 2023 (registration number CRD42023399126).

2.2. Search Strategy

Relevant published scientific articles focusing on PPC referral criteria for children with CHD were searched via PubMed (including Cochrane), Embase and Web of Science. The following combination of Medical Subject Headings (MeSH) terms was used in PubMed: Paediatric Palliative Care referral criteria AND (children with complex heart disease OR complex cardiac disease). The inclusion criteria were: studies in Portuguese and English published between 2013 and 2023, and age (birth to 18 years old). The query used in the Embase database was 'Paediatric Palliative Care'/exp AND 'children with complex heart disease'/exp. The inclusion criteria were: articles published since 2013, age (newborn, infant, child, preschool child, school child, adolescent) and language (Portuguese and English). The query used in the Web of Science database was: Paediatric Palliative Care (Title) AND children with complex heart disease (Title). The literature was narrowed to English and Portuguese language studies published within the last 10 years. In addition, reference lists of landmark studies and review articles on the subject were searched, in order to identify any relevant studies that might have been missed in the online searches performed.

2.3. Eligibility Criteria

Original articles, reviews (with or without meta-analyses), randomised trials, observational (including cohort and case-control), experimental and multicentric studies regarding the criteria for CHD referral to PPC care were considered eligible, whereas unpublished studies were excluded from the review.

Titles and abstracts were evaluated with respect to relevance and study design, according to the inclusion criteria. The complete papers were subsequently checked against the eligibility criteria. We excluded studies whose outcomes were not focused or explicit with respect to the referral criteria of children with CHD to PPC. Another exclusion criteria was the duplication of sources, i.e. articles where records were already equated or in which the author failed to give their personal opinion.

2.4. Ethical Issues

The principles of loyalty and respect for authors, in what concerns text integrity, were safeguarded through the inclusion of references. No excerpts were used out of context or with a different interpretation, to preserve the original meaning. Considering the study design, no approval by the Ethics Committee and/or the National Data Protection Committee was required.

3. Results

3.1. Characteristics of the studies included

The PubMed database query originated a total of 32 articles, of which 19 were excluded, based on the abstract, for not being specific to paediatric cardiology. Of the remaining articles, three were discarded for not having records that were not already being equated or in which the author failed to give their personal opinion. Thus, a total of 10 articles from the PubMed database were included.

The Embase database query originated a total of six articles, of which two were duplicated articles; three other articles were excluded for not being paediatric cardiology related. Therefore, no articles from Embase database were included.

The Web of Science query originated five articles, of which all were excluded, as three had already been retrieved using the previous databases, whereas the remaining two did not meet the pre-established inclusion criteria. Thus, no studies from Web of Science were included.

Of the 43 articles retrieved throughout the literature search conducted, as well as an additional three articles included through other sources, three duplicates were excluded and the remaining 43 screened. Of the latter, 28 articles were excluded based on title or abstract content and 15 were reviewed by full text. In total, 11 studies were considered eligible and included in the current systematic review, as shown in Figure 1.

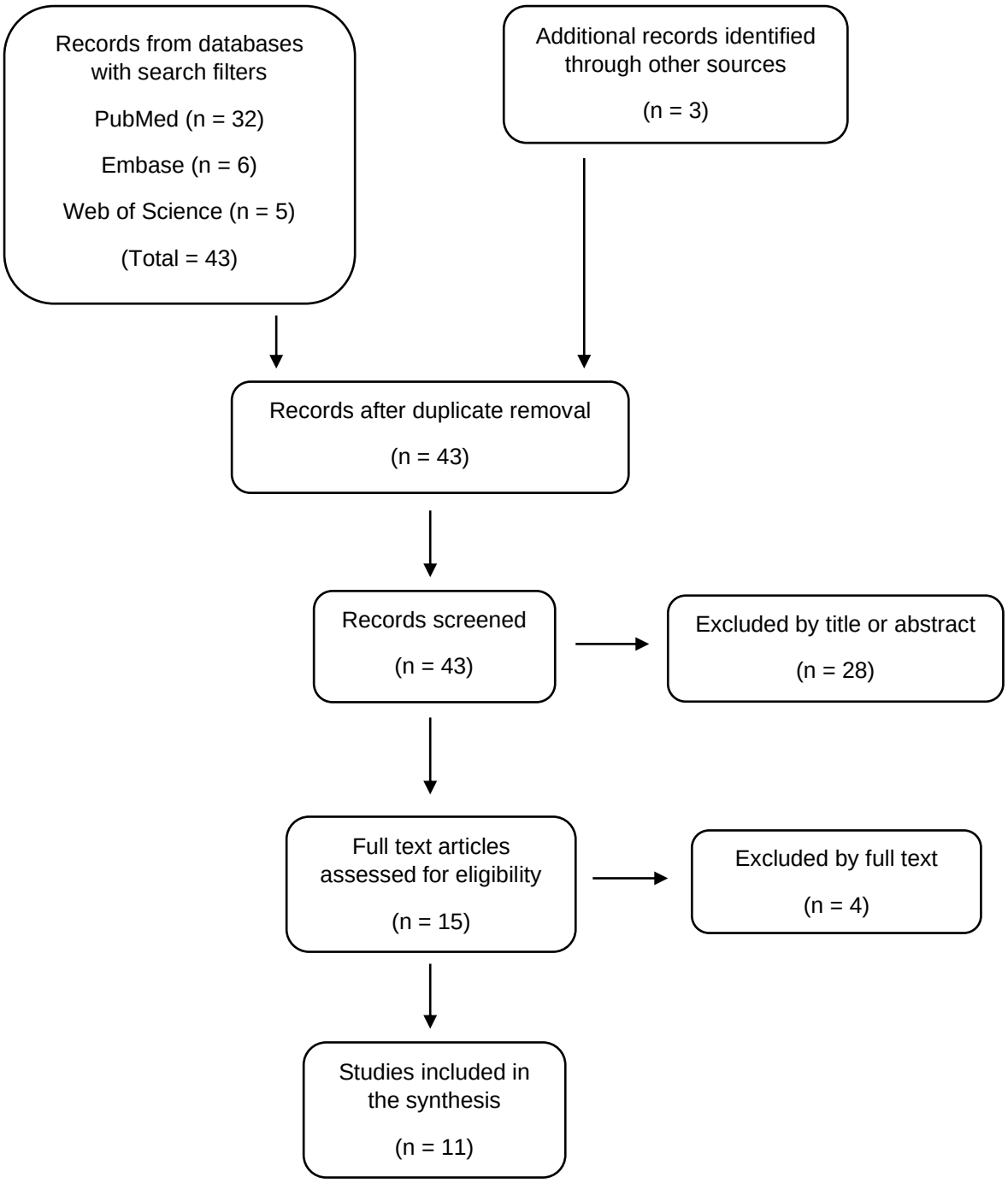

Figure 1 Flowchart of search strategy and selection of studies for inclusion in the systematic review: paediatric palliative care in children with complex heart disease – when to refer?

The main PPC referral criteria that each eligible study used to children suffering from CHD are summarized in Tables 1 and 2.

Table 1 Main Results of the studies analysed

Authors	Publication Year	Study Design	Main referral criteria
Hope, K. D. *et al*	2021	Review	• Mechanical circulatory support (ECMO/VAD) • Considered for heart transplant • Congenital heart disease and HF
Kaufman, B. D. *et al*	2019	Review	• Symptomatic HF • Consideration of mechanical circulatory support (ECMO/VAD) • ICD • Controversial/uncertain treatment pathways
Morell, E. *et al*	2019	Review	• SVHD • Congenital heart disease with associated genetic syndrome • Pulmonary vein stenosis • Pulmonary hypertension • Evaluation or listing for heart transplant • Need for mechanical circulatory support (ECMO/VAD) • Hospital admission length > 30 days or > 3 per year • Multiple cardiac surgeries or catheterisations (> 3 times in 1 year) • Mechanical ventilator support > 14 days • Not amenable to intervention and where curative treatment strategies have failed
Mazwi, M. L. *et al*	2016	Review	• SVHD • Congenital heart disease with associated genetic syndrome or comorbidities • Symptomatic HF or significant myocardial dysfunction • Being considered for heart transplant • Requiring mechanical circulatory support (ECMO/VAD) • Indication for an ICD. • Controversy about the appropriate intervention or uncertainty about eventual outcome
Kirk, R. *et al*	2014	Review	• ECMO or VAD support • Listed for heart transplant • Other potential end-stage HF
La Fay, C. *et al*	2021	Retrospective record review	• Congenital heart disease with great severity (Bethesda classification) • Congenital heart disease with moderate severity (Bethesda classification) and extra-cardiac anomalies • Cardiomyopathy and extra-cardiac anomalies
Marcus, K. L. *et al*	2018	Retrospective record review	**Congenital/structural disease** • SVHD • Pulmonary vein stenosis • Other – tetralogy of fallot; severe valve disease

			Acquired/non-structural disease • Cardiomyopathy • Pulmonary hypertension • Myocarditis **During the admission related to initial PPC consultation:** • Mechanical ventilator support • ECMO or VAD support • ≥ 1 cardiac arrest • Other – vasoactive medications
Delgado-Corcoran, C. *et al*	2020	Retrospective cohort	• SVHD • Cardiomyopathy • Myocarditis • Heart transplant • Underlying neurological and congenital/genetic diagnosis
Vemuri S. *et al*	2021	Cross-sectional Survey	• No further surgical options • SVHD • VAD • Comorbid genetic conditions • Symptomatic HF from myocardial dysfunction • Awaiting heart transplant
Balkin, E. M. *et al*	2018	Cross-sectional survey	• VAD • Heart transplant • ECMO • Pulmonary hypertension • Associated genetic diagnoses • SVHD • Pulmonary vein stenosis • Lung transplant • Extreme prematurity
Balkin, E. M. *et al*	2017	Cross-sectional survey	• Pulmonary vein stenosis • VAD • Heart transplant • ECMO

ECMO - Extracorporeal membrane oxygenation; **HF -** Heart failure; **ICD -** Implanted cardiac defibrillator; **SVHD** - Single ventricle heart defects; **VAD -** Ventricular assist device.

Table 2 Referral criteria for children with complex heart disease to paediatric palliative care and relative frequencies

Referral Criteria	Number of articles cited	Frequency
Congenital/structural heart disease		
SVHD	6	54.5% (6/11)
Pulmonary Vein Stenosis	4	36.4% (4/11)
With symptomatic HF	4	36.4% (4/11)
With great severity (Bethesda Classification)	1	9.1% (1/11)
Other	1	9.1% (1/11)
Acquired/non-structural heart disease		
With symptomatic HF or significant myocardial dysfunction	4	36.4% (4/11)
Pulmonary hypertension	3	27.3% (3/11)
Cardiomyopathy	2	18.2% (2/11)
Myocarditis	2	18.2% (2/11)
Advanced therapeutic strategies (Consideration)		
VAD	9	81.8% (9/11)
Heart transplant	8	72.7% (8/11)
ECMO	8	72.7% (8/11)
ICD	2	18.2% (2/11)
Other	2	18.2% (2/11)
Prolonged mechanical ventilatory support	1	9.1% (1/11)
Burden of treatment		
No further treatment options	2	18.2% (2/11)
Controversial/uncertain treatment pathways	2	18.2% (2/11)
Multiple cardiac surgeries or catheterizations	1	9.1% (1/11)
Multiple and/or prolonged hospitalizations	1	9.1% (1/11)
Associated Comorbidity		
Genetic syndromes / chromosomal anomalies	6	54.5% (6/11)
Other comorbidities (e.g. neurological)	2	18.2% (2/11)
Other	2	18.2% (2/11)

ECMO - Extracorporeal membrane oxygenation; **HF -** Heart failure; **ICD -** Implanted cardiac defibrillator; **SVHD** - Single ventricle heart defects; **VAD -** Ventricular assist device

An effort was made to systemise the best approach to these patients by gathering all available scientific evidence on current recommendations for referral children with CHD to PPC, since literature is not always consensual.

Hope, K. D. *et al* (2021)[43] conducted a literature review on the scope of PPC in the realm of paediatric HF, ventricular assist device (VAD) support and heart transplant. The authors reported that PPC may be especially beneficial in case of uncertainty about heart transplant or re-transplant, as well as patients on VAD support as a bridge to a decision or as destination therapy, and congenital HD and HF compounding their chronic healthcare needs.

Kaufman, B. D. *et al* (2019)[17] analysed several articles in order to examine the evolving role of PPC in supporting children with AHD. The main specific triggers for PPC consults proposed by the authors included symptomatic HF, consideration of mechanical circulatory support, implanted cardiac defibrillator (ICD), or controversial/uncertain treatment pathways. However, they considered, based on their experience, that early PPC involvement is indicated for children affected with CHD and being evaluated for VAD support or heart transplant, and also for patients with HF or AHD and uncertain outcomes.

In 2014, Kirk, R. *et al*[44] listed the guidelines for the management of paediatric HF from the International Society for Heart and Lung Transplantation, according to which PPC is recommended in all children with potentially end-stage HF, including patients receiving extracorporeal membrane oxygenation (ECMO), VAD support, or listed for transplant.

Morell, E. *et al* (2019)[45] carried out a 10-year review of the relevant literature about the intersection between PPC and paediatric cardiology in AHD. They defended the following PPC referral criteria: SVHD physiology, congenital HD with associated genetic syndrome, pulmonary vein stenosis, pulmonary hypertension, evaluation or listing for heart transplant, mechanical circulatory support, hospitalisation period longer than 30 days, multiple cardiac surgeries or catheterisations (more than three in one year), mechanical ventilator support for more than 14 days, more than three hospital admissions per year due to cardiac conditions (arrhythmia and HF) and not being amenable to intervention (curative strategies have failed).

The article of Mazwi, M. L. *et al* (2016)[26] aimed to broadly discuss the role of PPC in the management of children with critical congenital HD. The authors considered the following clinical scenarios as reasonable triggers for PPC involvement: any lesion related with SVHD physiology, congenital HD associated with a genetic syndrome or comorbidities, symptomatic HF or significant myocardial dysfunction, being considered for heart transplant, requiring mechanical circulatory support (ECMO and/or VAD), meeting an indication for an ICD, or controversy about the appropriate intervention or uncertainty about the outcome.

La Fay, C. *et al* (2021)[31] conducted a retrospective medical chart review involving six French PPC teams and all 18 the patients who were referred to them for AHD and had a PPC consultation in 2019, in order to describe the PPC role in those children. Six of these patients (33.33%) had cardiomyopathy and 12 (66.67%) had congenital HD (according to the Bethesda classification, three with moderate severity and extra-cardiac anomalies, and nine with great complexity). Additional extra-cardiac anomalies, including genetic syndromes, chromosomal disorders, and defects of other systems, were found in seven of 12 (58.33%) patients with congenital HD and five of six (83.33%) with cardiomyopathy, totalling 12 of 18 cases (66.67%). Their results suggest that patients with extra-cardiac anomalies are more likely to be referred to PPC teams. The conclusion that can be drawn from the data is that the main reasons for referral were congenital HD with great complexity, congenital HD with moderate severity and extra-cardiac anomalies, or cardiomyopathy with extra-cardiac anomalies.

Marcus, K. L. *et al* (2019)[10] conducted a retrospective single-institution medical record review of 201 patients with AHD for whom the PPC team was initially consulted between 2011 and 2016, for the purpose of examining features of PPC involvement. A total of 87% of these patients presented with congenital/structural HD (including SVHD (48%), Tetralogy of Fallot with pulmonary atresia/double outlet right ventricle/complete atrioventricular canal type (21%), pulmonary vein stenosis (13%), severe valve disease (6%) and other conditions (12%)). The remaining patients (13%) had acquired/non-structural HD (including cardiomyopathy (81%), pulmonary hypertension (11%) and myocarditis (7%)).

To capture the clinical circumstances leading up to the initial PPC consultation, features of the most recent hospitalization were recorded. As a reflection of lifetime disease burden, 70% of patients were mechanically ventilated upon admission, 76% were receiving vasoactive medications and 24% were being supported by ECMO or VAD. Regarding nonfatal cardiac arrests, 15% experienced one event during this admission, and 6% suffered two or more. The authors concluded that patients predominantly had congenital/structural HD, and most were receiving intensive therapies, such as mechanical ventilation or vasoactive medications, on the initial consultation (on average, one month passed until death since then).

Delgado-Corcoran, C. *et al* (2020)[46] conducted a retrospective cohort study in 1389 children with CHD admitted to the cardiac intensive care unit (CICU) from January 2014 to June 2017 in a university-affiliated tertiary care children's hospital, for the purpose of describing PPC consults in that population, applying Center to Advance Palliative Care (CAPC) criteria for PPC and determining the impact of this on EOL. These guidelines include, but are not limited to, SVHD, cardiomyopathy, myocarditis, cardiac transplant, and a combination of cardiac diagnosis with underlying neurological and congenital/genetic diagnosis.

These authors compared the characteristics of paediatric patients, with all types of CHD, treated in a dedicated CICU, who received a PPC consult, with patients who did not, having realized apparent missed referrals when CAPC guidelines were retrospectively applied. They concluded that those criteria were not widely used and their impact had not been reported.

Vemuri S. *et al* (2021)[25] performed a cross-sectional survey of 171 professionals (91 from paediatric cardiology and 86 from PPC), in order to explore their perspectives on the provision of PPC to children with CHD. An invitation and secure email link were sent to several associations and societies. The survey included close-ended questions with answer choices in 4-point Likert scales, and open-ended ones, having been open for approximately two months. The authors concluded that their study provides evidence-based guidance for integration and delivery of PPC to children suffering from CHD and the consensual criteria.

The clinicians agreed that children with no further surgical options, comorbid genetic disorders, diagnosis of SVHD, VAD *in situ*, symptomatic HF from myocardial dysfunction and/or awaiting heart transplant would benefit from PPC involvement. Although there was no agreement among cardiologists, no significant overall differences between these specialists and PPC professionals was found with respect to prolonged ICU stay, neonates with SVHD proceeding to surgery, and lack of consensus over the treatment plan. Neither group agreed on PPC involvement for a planned VAD, ICD or ECMO.

In 2018, Balkin, E. M. *et al* [39] performed a cross-sectional survey, retrospective over a 5-year period. An introductory letter and the survey were sent by e-mail, as well as two reminders, at monthly time intervals, in order to compare the perspectives of 183 paediatric cardiologists and 49 PPC physicians of one single centre, regarding PPC in paediatric HD.

Automatic consultations were reported for conditions such as ECMO, heart or lung transplant, extreme prematurity, trisomy 13 or 18, pulmonary vein stenosis and SVHD. No significant difference between the groups was found regarding the perceived extent of PPC involvement (often/always) for heart transplant (39%-53%), pulmonary hypertension (22%-42%), VAD (43%-53%), and ECMO patients (30%-35%). However, PPC physicians perceived greater PPC involvement (often/always) in the care of patients with SVHD (40% vs. 19%) and for CHD with associated genetic diagnoses (86% vs. 59%). In contrast, these physicians perceived a less frequent involvement in patients with pulmonary vein stenosis (23% vs. 41%). Both groups reported greatest PPC team involvement in patients with underlying genetic syndromes, SVHD, and in those who have received a heart transplant or VAD.

The study of Balkin, E. M. *et al* (2017)[11] was a cross-sectional survey of 547 paediatric cardiologists and 42 cardiac surgeons from 19 paediatric medical centres conducted for the

purpose of understanding their perspectives and perceived competence regarding PPC concepts and involvement. An introductory letter and the survey, including primarily close-ended questions with answer choices presented in Likert scales, were sent by e-mail, as well as a reminder, four months after the initial request, in order to solicit additional responses.

About a third of the respondents admitted that PPC occurred “often” or “always” at their institutions for children with pulmonary vein stenosis (37%), heart transplant (33%), VAD (35%), and ECMO (30%), but around 25% reported that PPC was “never” or “rarely” involved.

Through a consensus between the authors, we tried to create a workflow, for the purpose of assisting health professionals from both areas with the achievement of a correct, timely referral, in a simpler, more objective manner, as a guide for clinical practice. In this approach, a consideration for PPC consultation is initiated based on diagnosis, associated comorbidity, advanced therapeutic strategies, and/or burden of treatment.

3.2. Algorithm for referral to PPC

After analysing all the studies, we consider children with a cardiac condition involving SVHD physiology (e.g. HLHS and heterotaxy with single ventricle), pulmonary vein stenosis, pulmonary hypertension, symptomatic HF (excluding those caused by left-to-right shunt susceptible to surgical correction or percutaneous closure) or significant myocardial dysfunction have CHD and should be automatically referred to a PPC consultation, as well as children with associated trisomy 13 or 18, and patients receiving mechanical circulatory support (VAD/ECMO) or a heart transplant.

Because they are extremely pertinent, we also propose that the main trigger points for PCC used by Benini *et al*[40], in International Standards for Pediatric Palliative Care, for general paediatrics be included in this tool whenever children may have an aforementioned condition.

Additionally, the team may consult the PaPaS Scale[41] when:

1. The child has a cardiac condition involving other diagnoses or comorbidities (specially neurological);
2. An ICD implantation or other advanced therapeutic strategies (e.g. lung transplant, mechanical ventilator support, vasoactive medication) are being considered or applied;
3. There is a burden of care, due to caregiver exhaustion, or treatment (e.g. multiple cardiac surgeries or catheterizations, multiple and/or prolonged hospitalizations or mechanical ventilation, no further or uncertain treatment options), as well as other circumstances, such as nonfatal cardiac arrest and extreme prematurity;

Our management algorithm for referral children with CHD to PPC is shown in figure 2.

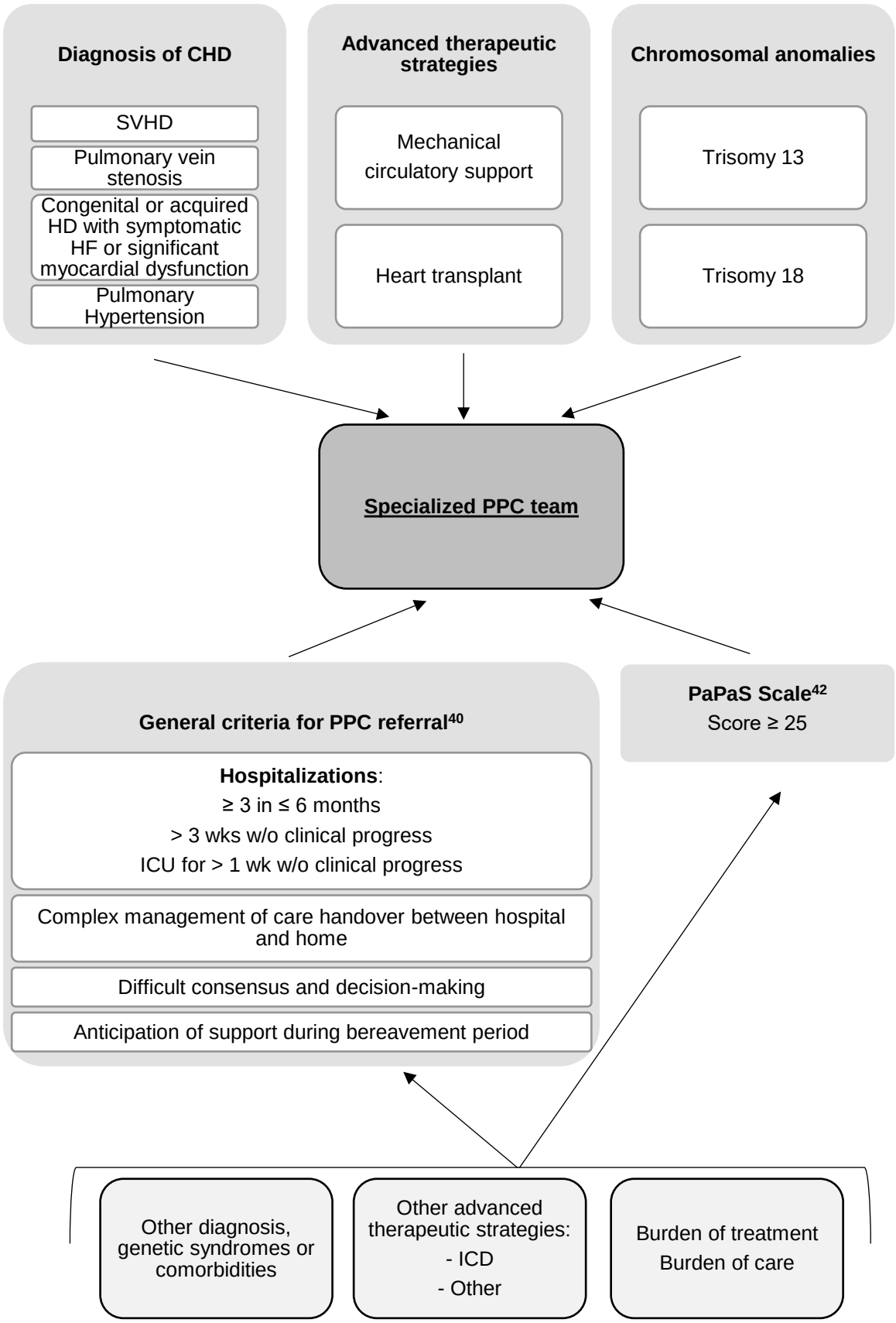

Figure 2 Proposed management algorithm for referral children with complex heart disease to paediatric palliative care. **HD** – Heart disease; **HF** - Heart failure; **ICD -** Implanted cardiac defibrillator; **PaPaS -** Paediatric Palliative Screening; **PPC** - Paediatric palliative care; **SVHD** Single ventricle heart defects; **Wk** – week; **Wks** – weeks**; w/o** –without.

4. Discussion

CHD is the leading cause of non-accidental death in childhood[14,36] and neonatal morbidity (affecting 1.35 million neonates annually worldwide), causing ongoing morbidity throughout the course of a child's life[6]. Death is more likely to occur in the first year of life[14,42,47,48], commonly in high-acuity settings following withdrawal of invasive support in intensive therapies[10,49].

The retrospective cohort study of Feudtner C. *et al* (2001)[50] analysed the trends in the pattern of deaths attributable to paediatric CCC over a 20-year period and showed that cardiovascular diagnoses were the main causes of death in those children (29%), reinforcing the huge impact of cardiac conditions.

Lacerda, A. J. *et al* (2017)[51] conducted a 25-year (1987-2011) study on paediatric deaths in Portugal, showing that CHD was the first cause of death in the first year of age, and counts to 24.4% in older children.

Lacerda, A. J. *et al* (2019)[48] performed another 5-year (2011-2015) study in portuguese hospitals, which showed that the absolute number of admissions of paediatric patients has decreased, although the admissions of children with CCC and PPC needs have risen proportionally, with longer episodes and higher mortality than those observed in other patients (trends intensified in the presence of two or more CCC categories). Cardiovascular conditions were found to be the third most predominant CCC category (15.4%), with the third highest mortality rate (3.3%).

Despite the observed increase in life expectancy brought about by advanced therapies, the QoL of patients was found to have worsened, affecting both the children and their families[23,27], with parents reporting significant suffering of their children at the EOL and only being able to realise the severity of the prognosis and impending death within the last 24-48 hours of the children's life.[14,36]

A study of bereaved parents of children with AHD, 47% reported that their child experienced significant suffering in the month prior to death, with 71% describing health-related QoL as "poor" or "fair"[36]. Significant disparities were also observed between parents and physicians in what regards symptom burden, perceived suffering and expectations about prognosis and QoL at time of diagnosis, which suggests the existence of gaps in parent–physician communication. Variable communication also occurs in family counselling, particularly concerning the discussion of diagnoses[11].

Nogueira A. *et al* (2022)[52] performed a retrospective cohort study with the aim of describing the last year of life of 73 children who died with CCC in a tertiary portuguese hospital between January 2016 and December 2020, among which 15% with CHD. They found out that PPC

reduces the use of invasive therapies and procedures (e.g. resuscitation methods, endotracheal intubation, central venous catheter placement, mechanical invasive ventilation and transfusion of blood products), decision to limit treatment and deaths in ICU. However, EOL care in general remains one of the most neglected areas in Portugal's health system.

Unfortunately, information and literature on the treatment approach for children with CHD and the incorporation of PPC is still lacking, as no specific guidelines are available for early referral, leading to a positive impact in child's QoL, with an individualized care plan to address theirs needs and wishes.

After analysing the studies included, the authors found some consensual criteria, that must be immediately considered for PPC referral, which were:

1. SVHD physiology (e.g. HLHS and heterotaxy with single ventricle);
2. Pulmonary vein stenosis;
3. Symptomatic HF (excluding those caused by left-to-right shunt susceptible to surgical correction or percutaneous closure) or significant myocardial dysfunction;
4. Pulmonary hypertension;
5. Trisomy 13 or 18;
6. Children receiving mechanical circulatory support (VAD/ECMO) or a heart transplant.

Regarding genetic and chromosomal disorders may increase complexity. Patients with trisomy 13 (Patau syndrome) or 18 (Edwards syndrome) have severe comorbidities and poor prognosis with > 90% of the affected infants dying by age one year. Despite poor overall survival, given the multi-systemic nature of the disease, the presence of CHD may not impact it. In fact, cardiac operations may be beneficial in some groups. Thus, the care for these patients and families requires a balanced multidisciplinary approach, including PPC teams[21].

The literature indicates that the median survival time of a transplanted heart is 12-20 years, depending on the recipient's age, what can subject them to numerous challenges of worsening HF, becoming too sick to be able to survive a surgery, and decisions about re-transplantation. Moreover, mechanical circulatory support (ECMO/VAD), despite its benefits, also carries potentially serious complications, making the decision-making process harder. All this justifies the fact that they are in direct referencing[43].

Although generalised criteria for referral to PPC have been defined, reflection based on the diagnosis-related group may raise doubts, particularly when invasive curative procedures are available. Despite the scarcity of robust literature, general criteria have been found to exist. In this sense, the authors have established an algorithm for referral to PPC and stressed the

importance of timely referral for avoiding unnecessary procedures and ensuring the adoption of a treatment approach suited to the needs of the children and their families.

Therefore, we made a management algorithm, adapted to our population's features and fragilities, based on our systematized results, emphasizing the multidisciplinary approach and advance care planning importance to early meet their needs and improve their outcomes. Families would also be able to benefit from this approach, as it would address their needs and allow them to better identify their rights.

In ideal terms, the model would not only be easy to access and implement in paediatric cardiology services, but also draw the attention of specialists to the importance of PPC and referral to these services, especially considering that the referral rates reported in the literature are still significantly low.

4.1. Limitations

This systematic review considered all the published evidence that included referral criteria for children with CHD, having also covered the opinions of authorities and/or reports of expert panels. This may represent a limitation, as the latter data may have a higher degree of subjectivity. Due to the response rate, it is likely that most responses were provided by professionals with greater interest in the subject, in which case the most contradictory opinions may not be represented in the sample (lack of inclusion), limiting the generalizability of the study (sampling bias), which may contribute to an over-estimation of perceived knowledge and competence in certain areas. Also, evidence base for many of the recommendations was found to be low, due to the lack of trials and information in children.

Some retrospective cohort studies are limited by referring to a single centre and having a small sample, which restricts the diversity of patient characteristics and does not allow for an accurate comparison with other centres. Since the information was obtained from medical and administrative records, documentation may be lacking. On the other hand, two articles failed to include specific referral criteria, as only the population was described. In these cases, the authors transposed the reasons they considered relevant for receiving PPC.

5. Conclusion

CHD is a major cause of morbidity and mortality in children, causing huge suffering and considerably affecting the QoL of patients. Given the scarcity of literature and the low level of evidence of the available data, we believe it is urgent to establish clear guidelines in paediatric cardiology, such as to increase awareness of the importance of early referral to PPC.

This population requires a patient-oriented healthcare model designed to meet their needs. In this sense, we gathered all the relevant data about referral criteria for PPC and sought to develop a cohesive workflow of PPC management for the correct, early, timely identification, screening and assessment of children affected with CHD, as a clinical practice guideline. The triggers to forward them to a PPC consultation should be systematic enough to improve QoL and quality of care.

Next author's aim is to implement this recommendation in pediatric cardiology services, particularly in Hospital Pediátrico do Centro Hospitalar e Universitário de Coimbra, as an approach protocol.

Future studies are required in order to analyse the applicability of the proposed algorithm in Paediatric Cardiology settings and to assess the impact of timely referral of children with CHD on QoL, symptomatic treatment and alignment between care plans and the wishes and choices of patients and their families.

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7. Conflict of interest

The authors declare that there were no conflicts of interest in writing this article.

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Appendix I – Portuguese version of PaPaS Scale[42]

Escala PaPaS			**Número:**
Domínio e Números de Itens	**Item**	**Característica**	**Pontuação**
Domínio 1	**Trajetória da doença e impacto nas atividades de vida diária da criança/jovem**		
1.1	Trajetória da doença e influência nas atividades diárias da criança/jovem (comparação com a atividade de base própria da criança/jovem nas últimas 4 semanas)	-Estável. -Deterioração lenta sem influência nas atividades diárias. -Instável e com influência nas atividades diárias e restriçãodas mesmas. - Deterioração significativa com restrição grave das atividades diárias.	0 ☐ 1 ☐ 2 ☐ 4 ☐
1.2	Aumento do nº de internamentos hospitalares (> 50% em 3 meses, comparado com períodos anteriores)	Não Sim	0 ☐ 3 ☐
Domínio 2	**Resultado esperado do tratamento da doença e efeitos secundários associados**		
2.1	Tratamento da doença (não significa tratamento de complicações relacionadas com a doença: ex dor, dispneia ou fadiga)	-… é curativo. -… controlo da doença e prolonga a vida com boa qualidadede vida. -… não cura nem controla a doença, mas tem um efeito positivo na qualidade de vida. -… não controla a doença e não tem efeito na qualidade de vida.	0 ☐ 1 ☐ 2 ☐ 4 ☐
2.2	Efeitos secundários do tratamento (incluindo influência na família e no doente, por ex: internamentos na perspetiva do doente ou família)	-Nenhum ou ligeiros -Ligeiros -Moderados -Graves	0 ☐ 1 ☐ 2 ☐ 4 ☐
Domínio 3	**Sinais/Sintomas e problemas**		
3.1	Intensidade de sinais/sintomas e/ou dificuldade no controlo destes (últimas 4 semanas)	-Assintomático -O(s) sinal(s)/sintoma(s) é(são) ligeiros e fáceis de controlar -Qualquer sinal/sintoma é moderado e controlável -Qualquer sinal/sintoma é grave ou difícil de controlar (hospitalização não planeada ou visitas em ambulatório, sinais/sintomas em crise)	0 ☐ 1 ☐ 2 ☐ 4 ☐
3.2	Distúrbios psicológicos (stress) do doente relacionados com os sinais/sintomas	-Ausente -Ligeiro -Moderado -Significativo (grave)	0 ☐ 1 ☐ 2 ☐ 4 ☐
3.3	Distúrbios psicológicos (stress) dos pais ou família relacionados com os sinais/sintomas e sofrimento da criança	-Ausente -Ligeiro -Moderado -Significativo (grave)	0 ☐ 1 ☐ 2 ☐ 4 ☐
Domínio 4	**Preferências/necessidades do doente ou pais /Preferências do profissional de saúde**		
4.1	O doente/Os pais deseja(m) receber cuidados paliativos ou expressa(m) necessidades equivalentes aos cuidados paliativos	-Não -Sim	0 ☐ por favor, responder a 4.2 4☐ não responder a 4.2
4.2	Você ou a sua equipa sente(m) que este doentebeneficiaria dos cuidados paliativos	-Não -Sim	0 ☐ 4 ☐
Domínio 5	**Esperança de vida**		
5.1	Estimativa da esperança de vida	-Vários anos -Entre meses a 1-2 anos -Entre semanas e meses -Entre dias e semanas	0 ☐ por favor, responder a 5.2 1 ☐ por favor, responder a 5.2 3 ☐ não responder a 5.2 4 ☐ não responder a 5.2
5.2	"Ficaria surpreendido se esta criança morresse repentinamente no prazo de 6 meses?"	-Sim -Não	0 ☐ 2 ☐
		Pontuação total:	