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# Patients requiring pediatric palliative care for advanced heart disease in France: a descriptive study

Short title: Patients requiring pediatric palliative care for advanced heart disease

C. La Fay[a*], P. Le Moine[b], L. Blanc[c], W. Abou Chahla[d], N. Phan Hoang[e], E. Vial-Cholley[e], N. Cojean[f] , A. Suc[g], P. Saultier[h], E. Aries[i], C. Ovaert[i], G. Revon-Rivière[a] on behalf of Fédération des Equipes Ressources Régionales de Soins Palliatifs Pédiatriques (ERRSPP).

[a]ERRSPP PACA, Timone Children's Hospital, AP-HM, 264 rue Saint-Pierre, 13385 Marseille, France;

[b]ERRSPP La BRISE, 2 avenue Foch, 29609 Brest, France;

[c]ERRSPP Poitou-Charente, 2 rue de la Milétrie 86021 Poitiers, France;

[d]ERRSPP Nord Pas-de-Calais, Hôpital Albert Calmette, 59037 Lille, France;

[e]ERRSPP IHOP, 1 place Joseph Renaut, 69008 Lyon, France;

[f]ERRSPP Alsace CHU Hautepierre, Avenue Molière, 67098, Strasbourg;

[g]Enfant-do, Children's hospital, (CHU), TSA 70034, 31059, Toulouse, France;

[h]Aix Marseille Univ, INSERM, INRAe, C2VN, Marseille, France;

[i]Department of Pediatric and Congenital Cardiology, Timone Enfant, AP-HM, 264 rue Saint-Pierre, 13385 Marseille, France.

***Corresponding author:** Charlotte LA FAY,

Timone Children's Hospital, 264 rue Saint-Pierre, 13385 Marseille, France

[charlotte.la-fay@ap-hm.fr],04 91 38 81 37.

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**ABSTRACT**

**INTRODUCTION:** Pediatric palliative care (PPC) teams address unmet needs and improve the quality of life of patients with life-limiting conditions across pediatric subspecialties. However, little is known about the timing, reasons, and nature of PPC team interventions in advanced heart diseases (AHD).

**OBJECTIVES:** Here we describe how, when, and why PPC teams interact with referred teams of children suffering from AHD.

**METHODS:** We conducted a retrospective nationwide survey among PPC teams in France. All patients referred to participating PPC teams for a cardiologic disease in 2019 were studied.

**RESULTS:** Among six PPC teams, 18 patients with AHD had a PPC consultation in 2019. Six of these patients had cardiomyopathy and 12 had congenital heart disease (CHD). The median age at referral was 0.9 months for CHD and 72 months for cardiomyopathy. An antenatal diagnosis had been made for six families with CHD, and two of them were referred to PPC before birth allowing for a prenatal palliative care plan. The main reason for referral was ethical considerations (50%) followed by organization for home-based palliative care (28%). PPC teams participated in ethical discussions when asked to but also provided family support (12/18), home-based PPC (9/18), coordination of care (5/18), support of the referred team (4/18), and symptoms management (3/18)

**CONCLUSION:** The main reason for referral to PPC was ethical considerations, but PPC interventions followed a holistic model of care. Prospective outcomes measurement and partnerships should be further developed.

**Keywords:** cardiology; pediatric; palliative care; advanced heart disease;

## 1. INTRODUCTION

Advanced heart diseases (AHD) are mainly caused by congenital heart disease (CHD) and cardiomyopathy [1-3]. CHD has a prevalence of 7 cases per 1,000 live births and accounts for nearly one third of major congenital anomalies [1]. Cardiomyopathy has an incidence of 1 per 100,000 children per year and is the leading indication for heart transplantation during childhood [4]. These conditions are responsible for impaired health-related quality of life (QOL), poorer school performance, less involvement in school activities, and physical impairment [5,6]. Although treatment for children with AHD mainly aims for total correction, mortality remains around 10% after the first congenital surgery [7]. Moreover, studies have shown that there is a need for improvement in communication, QOL, and advance care planning [8,9].

Pediatric palliative care (PPC) is the active total care of the child's body, mind, and spirit, and also involves giving support to the family [10]. In France, PPC teams were developed in each of the 22 metropolitan regions over the past decade. These teams foster implementation of PPC for children and teenagers within existing pediatric and adult facilities [11,12].

PPC has been shown to improve QOL, reduce the burden of disease, provide decision-making support, and increase communication about goals of care [13]. In pediatric life-limiting conditions, patients who received PPC spent fewer days in hospital, had fewer invasive interventions, and were less likely to die in intensive care units [14]. In a population with single-ventricle disease, a randomized controlled trial showed lower maternal anxiety, improved communication, and better family

relationships in the early PPC intervention group [15]. However, PPC remains underused in non-cancer pediatric specialties [13,16,17]. In a recent multicenter study with 515 patients, only 8% of PPC consultations were dedicated to cardiovascular diseases [17].

Early involvement of palliative care teams for patients with AHD is supported by several authors [18]. Mazwi et al. suggested indications for PPC consultations: CHD associated with genetic syndromes or comorbidities, any CHD involving single ventricle, symptomatic heart failure, and patients considered for cardiac transplantation [19]. Systematic referral to PPC teams based on such indicators would improve PPC access with respect to professional goals of care and parental hope [16].

In France, a registry on cardiologic palliative situations does not exist. Data on this population are scarce. Hence, we performed a preliminary investigation, by asking PPC teams to describe their interventions among patients with AHD.

## 2. **METHODS**

### *2.1 Study design and patients*

We conducted a retrospective chart review study of all patients suffering from a cardiologic life-limiting condition who were referred to the participating PPC teams in 2019 in France.

### *2.2 Data source*

A pediatric cardiology task force was formed within the French Federation of PPC teams (*Fédération des Equipes Ressources de Soins Palliatifs Pédiatriques*) [11] and participants agreed to become study coordinators for their own team. They reviewed medical charts of patients at their sites and sent a clinical description of the cases to the principal investigator, who compiled an anonymous master database.

Data were collected retrospectively from December 2019 to January 2020.

*2.3 Variables*

The following variables were collected: cardiac diagnosis (CHDs were described according to the Bethesda classification [20]), associated syndrome or disease, antenatal diagnosis, age at PPC referral, age at death, and length of follow-up. Follow-up started on the day of the initial PPC referral and ended when the patient died or on December 31, 2019 if the patients were alive at the time of data collection (i.e., maximum length of follow-up was 12 months).

The patient's age at the time of the PPC team intervention was categorized as: antenatal, neonatal (less than 1 month after birth), first year of life (1 month to 1 year) or more than 1 year old.

The reason for PPC team referral was defined as the main request stated in the clinical description and cross-checked by two authors (CLF, GRR). It was categorized into six groups as follows:

**Support in ethical considerations** was defined as support in discussions on advance care planning, therapy limitations (i.e., decision not to operate, do-not-resuscitate orders, interruption of a life-supportive intervention). These interventions did not necessarily imply meeting with the patient or their family.

**Home-based PPC** was defined as any support to an outpatient project. This included developing a care plan, training of general practitioners and home nurses, anticipation of medication needs, and home consultations by PPC team. This intervention typically implied a meeting with the patient and their family.

**Family support** was defined as any kind of supporting relationship with siblings, parents, or grandparents. This included discussions on advance care planning, patient and family education on distressing symptoms, psychological support, and bereavement support.

**Symptoms relief** was defined as clinical evaluation and prescription or help in prescription related to pain and symptom management.

**Coordination of care** was defined as communication between health-care providers, social workers, and teachers.

**Support of the referring team** was defined as interventions directed toward the referring team including help in defining ethical issues, solving communication issues, providing empathy, and encouraging self-preservation.

### *2.4 Statistical and graphical analysis*

Descriptive statistics (proportions, means, and medians) were used to describe patient characteristics, the reasons for PPC team referral, and the PPC team interventions.

### *2.5 Ethical statement*

Protocols for chart reviewing and medical data de-identification were reviewed by an independent ethics board of the French Federation of PPC teams (*Fédération des Equipes Ressources de Soins Palliatifs Pédatrique*) scientific committee and approved on November 22, 2019.

## 3. RESULTS

### *3.1 Patient characteristics.*

Six French PPC teams (Toulouse, Lille, Lyon, Marseille, Brest, and Strasbourg) shared their data of 18 patients whose characteristics are described in **Table 1**.

All patients who were referred to participant PPC teams in 2019 were reported here.

Six patients had a cardiomyopathy and 12 patients had a CHD. According to the Bethesda classification, nine of 12 patients had CHD of great complexity and three of 12 had CHD of moderate severity. All three patients with CHD of moderate severity had additional extra-cardiac anomalies.

In total, 12 of 18 patients had additional extra-cardiac anomalies. This included genetic syndromes, chromosomal anomalies, and anomalies of other systems.
Additional extra-cardiac anomalies were found in seven of 12 patients with CHD and five of six patients with cardiomyopathy.

### *3.2 Age at PPC referral*

The children's age at the initial PPC referral and indications on antenatal diagnosis are presented in **Table 1 and Figure 1**.

The median age at initial PPC referral was 4 months (range: 0–192 months): 0.9 months (range: 0–42 months) for patients presenting with CHD and 72 months (range: 0.3–192 months) for cardiomyopathy.

Half of the patients with CHD were diagnosed prenatally and the median age at PPC referral for these patients was 0.3 months (range: 0–36 months) compared with 2.3 months (range: 0.16–42 months) for patients with CHD diagnosed after birth. For two of these prenatally diagnosed patients, the PPC team was involved before birth.

*3.3 Length of follow-up.*

The length of follow-up is presented in Table 1. Overall median length of follow-up was 3 months (range: 2 days–12 months). While some patients had very short follow-up (four patients referred in the neonatal or antenatal period had 15 days or less of follow-up), most of them (14/18) had more than 1 month of follow-up. PPC teams were sometimes requested when patients were in a critical situation but the lifespan of the patient was finally longer than expected.

*3.4 Reasons for PPC team referral and interventions*

The first request for a PPC intervention came from cardiology teams in most cases (10/18), sometimes from neonatology teams or intensive care (4/18 and 3/18, respectively), and rarely from obstetrician teams (1/18). The main reason for PPC team referral was ethical considerations followed by organization of home-based PPC, family support, and symptom relief. Interventions were notably different from the reason for referral (**Figure 2**).

The interventions of PPC teams by frequency included family support (12/18), support in ethical considerations (11/18), home-based PPC (9/18), coordination of care (5/18), support of the referring team (4/18), and symptom relief (3/18).

Among the interventions that included support in ethical considerations, a discussion of therapy limitations took place in all cases (11/11).

Only one family received bereavement support. Coordination of care involved hospital-at-home services (3/5), district hospitals (2/5), and to a lesser extent home nurses, general practitioners, social workers, and teaching structures.

## 4. **DISCUSSION**

To our knowledge, this is the first clinical description of PPC involvement for patients diagnosed with AHD in France. These patients were cared for and their relatives supported by a collaborative effort of pediatric cardiology teams and PPC teams.

The population we described includes patients with CHD (2/3) and cardiomyopathy (1/3). The indications for PPC in France appear to be similar to other countries. In a recent retrospective study of PPC referral for 201 patients, Marcus et al. described 87% of CHD cases, 13% of cardiomyopathy cases, and 1% pulmonary hypertension cases. It is noteworthy that no patients with pulmonary hypertension were present in our cohort despite supporting literature for palliative care for this condition in both adult and pediatric settings [21]. Our results suggest that patients with extra-cardiac anomalies are more likely to be referred to PPC teams. Extra-cardiac anomalies (i.e., genetic syndromes, chromosomal anomalies, or anomalies of other systems) were found in 67% of the patients of our cohort in contrast to a frequency of 20–30% of children with AHD reported in the literature [1, 22]. Every patient with CHD of moderate severity (group 2) referred to the PPC team presented with an associated

congenital anomaly. This is consistent with previous studies of patients with CHD [16]. Extra-cardiac anomalies were also more frequent in patients with cardiomyopathy referred to PPC teams in comparison with larger series [23, 24]. This may be explained by an increased morbidity but may also reflect ethical issues about delivering intensive cardiologic treatment to vulnerable patients and concerns about their QOL [25, 26].

PPC teams were involved in the early course of the disease including some cases of prenatal PPC referral. We found early PPC involvement among CHD patients and earlier referrals in the case of prenatal diagnosis. Several studies state that PPC should be introduced prenatally in order to establish a longitudinal relationship and to avoid having the first PPC consultation during crisis situations [15, 27]. Early PPC involvement may help families to face complex decisions and avoid burdensome interventions [19].

However, considering the significant rate of poor outcome for patients with AHD, the number of patients referred to PPC teams in our study seems to be relatively low. Indeed, the infant mortality rate for all cases of CHD combined is 6.4% in France and more than 30 % of patients with cardiomyopathy die or undergo heart transplantation 1 year after diagnosis [1,2]. Moreover, patients with better outcome should also benefit from PPC interventions as they have a chronic condition with potentially repeated hospitalizations and surgical interventions.

We report more than 1 month of follow-up by PPC teams for 14 of 18 patients with AHD. There is evidence supporting the fact that the length of PPC follow-up

influences patient outcomes [8, 28] and that PPC teams should be introduced as soon as possible in order to improve QOL.

Support in ethical considerations and home-based PPC were the most frequent reasons for referral to the PPC team. These items define the core needs of cardiology teams. This is consistent with the findings of Marcus et al. on the indications for PPC consultation in AHD patients. In their retrospective study including 201 patients, discussions about the goals of care and provision of psychosocial support were far more frequent than symptom management, QOL, and coordination of care [18]. In France, this may be strengthened by the French law that allows for limitations in therapy when decided by the physician in charge, and after a collegial approach including the physician, an external consulting physician (i.e., who is not in charge of the patient), and members of the care team [29]. PPC physicians are frequently asked to participate as external consulting physicians or as legal experts.

Surprisingly, symptom management at the end of life was not considered an unmet need for the referring teams. This is noteworthy since a survey among pediatric cardiologists showed that more than half had no or only minimal competency with patient needs such as thirst, fatigue, gastrointestinal distress, and psychological distress [30]. This finding emphasizes the need for specialized PPC referral among children with AHD.

PPC teams implemented a holistic model of care even when they were called to resolve specific issues only. Although support in ethical considerations was systematically addressed when needed, PPC intervention simultaneously involved other dimensions of palliative care. As pediatric care specialists, we would advocate that family support, coordination of care, and support of the referring teams may

improve the patients' disease course through health-care services and may lower the distress of professionals caring for these patients.

Limitations of the current study include the low response rate. Only six of the 22 French regional PPC teams participated while a pediatric cardiology center exists in most regions of the country. Thus, this may not be fully representative of the whole country, although our findings with six different teams may offer a proper overview of the interactions between PPC teams and cardiology teams.

## 2. CONCLUSION

The collaboration between PPC teams and cardiologists appears to be feasible and is already in place in France. The development of clinical practice guidelines and common research goals should be pursued in order to strengthen collaboration and include more in-need patients with AHD.

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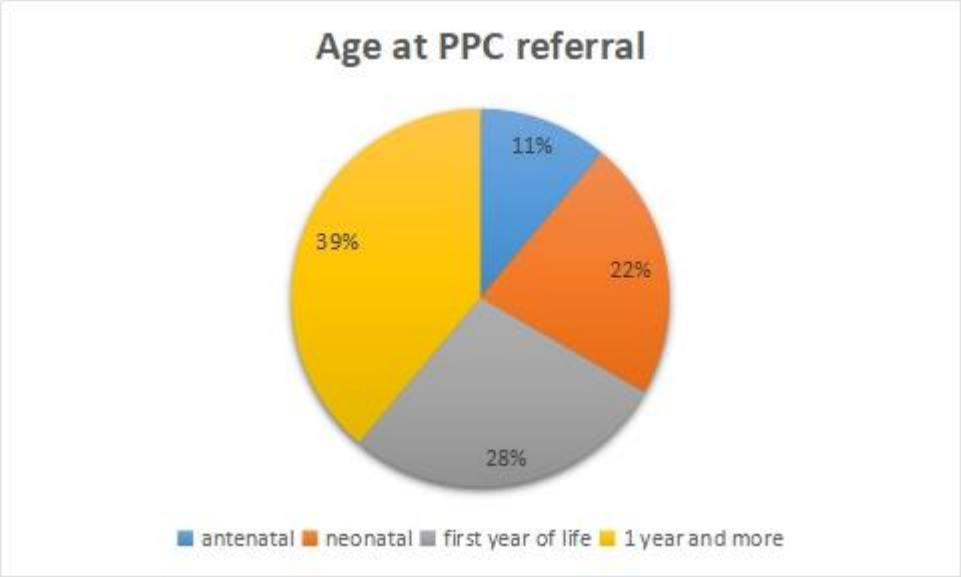
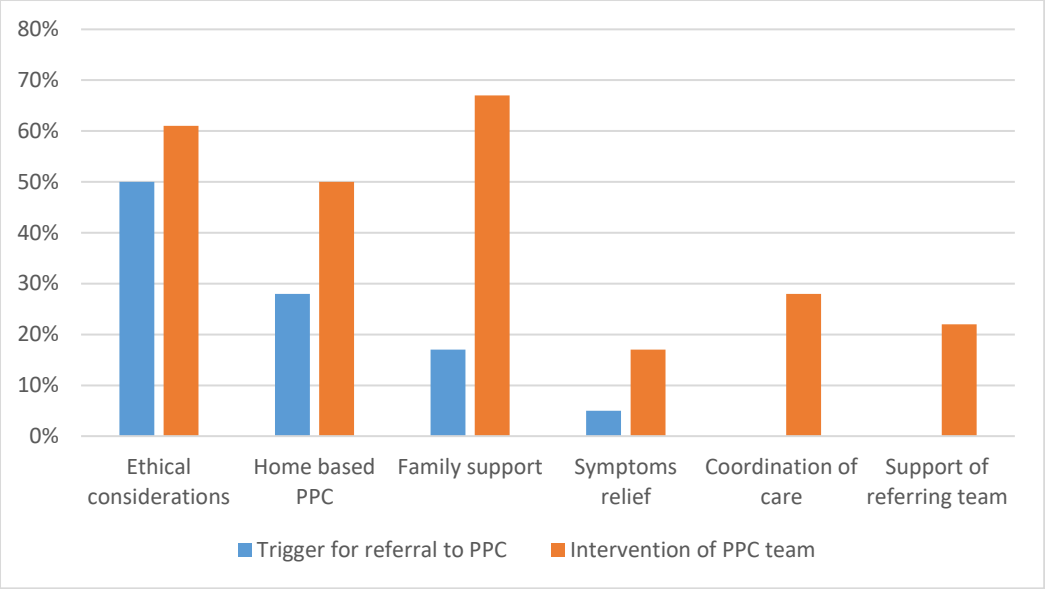
Figures:

**Figure 1**. Age at first pediatric palliative care (PPC) referral.

**Figure 2**. Reason for pediatric palliative care (PPC) team referral and intervention of PPC teams (*n*=18, %)


Age at PPC referral
11%
22%
28%
39%
antenatal
neonatal
first year of life
1 year and more


80%
70%
60%
50%
40%
30%
20%
10%
0%
Ethical considerations
Home based PPC
Family support
Symptoms relief
Coordination of care
Support of referring team
Trigger for referral to PPC
Intervention of PPC team

| | Cardiologic condition | Additional extra-cardiac anomalies | Antenatal diagnosis (Yes/no) | Age at PPC referral (months) | Length of follow-up, status |
|---|---|---|---|---|---|
| 1 | Dilated cardiomyopathy | Agenesis of the corpus callosum | No | 5.5 | 2 months, alive |
| 2 | Dilated cardiomyopathy | Friedreich's ataxia | No | 168 | 1 month, alive |
| 3 | Dilated cardiomyopathy | Doose syndrome | No | 192 | 1 month, deceased |
| 4 | Dilated cardiomyopathy | - | No | 9.5 | 3 months, deceased |
| 5 | Dilated cardiomyopathy, Heart transplant and graft rejection | 1p36 deletion syndrome | No | 108 | 12 months, alive |
| 6 | Restrictive cardiomyopathy | Chronic intestinal pseudo-obstruction | No | 36 | 4 months, deceased |
| 7 | Atrial septal defect and ventricular septal defect | Edwards' syndrome | Yes | At birth | >12 months, alive |
| 8 | Common atrioventricular canal | Down syndrome | No | 42 | 8 months, alive |
| 9 | Common atrioventricular canal | CHARGE syndrome | No | 0.3 | 2 days, deceased |
| 10 | Single ventricle with transposition | - | Yes | 1.3 | 3 months, deceased |
| 11 | Single ventricle with transposition and total abnormal pulmonary venous return | - | Yes | 0.5 | 8 months, deceased |
| 12 | Single ventricle, pulmonary atresia, interatrial communication | Ivemark syndrome | Yes | 0.1 | 0.5 month, deceased |
| 13 | Hypoplastic left heart and total abnormal pulmonary venous return | - | Yes | 36 | >12 months, alive |
| 14 | Hypoplastic right heart, pulmonary atresia and interatrial communication | - | No | 15 | 5 months, alive |

**Table 1.**

| | | | | | |
|---|---|---|---|---|---|
| 15 | Left congenital obstructive heart defects | - | Yes | At birth | 13 days, deceased |
| 16 | Pulmonary atresia with ventricular septal defect | Down syndrome | No | 1.5 | 1 month, deceased |
| 17 | Pulmonary atresia with ventricular septal defect | Di-George syndrome | No | 3 | 6 months, alive |
| 18 | Transposition of great arteries | CHARGE syndrome | No | 0.2 | 12 days, deceased |

**Patient characteristics**

PPC: pediatric palliative care