



## **Current practice of care for adolescent and adult patients after Fontan surgery in Poland**

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## **Current practice of care for adolescent and adult patients after Fontan surgery in Poland**

**Short title:** Practice of care for patients after Fontan surgery in Poland

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### **WHAT'S NEW?**

For the first time, the practice of care for patients after Fontan operation in Poland was systematically evaluated. In general, the frequency of routine visits and the assessment of basic cardiovascular status were consistent with the current guidelines in all centers. However, it was

shown that important surveillance tests in Fontan patients that are necessary to identify the early stages of associated complications are rarely used. The practice for care of adolescent Fontan patients differed from that of adults on many levels, including the frequency of routine visits and the proposed diagnostic assessment. As the variability in the surveillance testing regimen was shown in all participating centers, it was proven that there is no unified surveillance approach for Fontan patients in Poland. Our study emphasizes the urgent need to improve the organization of care for Fontan patients in Poland, which should be achieved with the support of the national healthcare service.

## **ABSTRACT**

**Background:** The growing number of adults patients after the Fontan operation requires regular surveillance tests in the specialized centers.

**Aims:** Evaluation of current practice of care for Fontan patients in Poland based on a multicenter survey.

**Methods:** Eight centers were included in the study-5 adult congenital heart disease (ACHD) and 3 pediatric centers for adolescents. To aim for a comparison between the centers and facilitate the interpretation of the results, the Fontan Surveillance Score (FSS) was developed. The higher score is consistent with better care, with a maximum of 19 points.

**Results:** The number of 398 Fontan patients (243 adults and 155 adolescents [age 14-18 years]) was included in the study. The median FSS was 13 points with variability between the centers (interquartile range 7–14 points). Centers providing continuous care from the pediatric period until 18 years of age achieved a higher FSS compared to ACHD centers (median: 14 points vs. 12 points;  $P < 0.001$ ). Most of the patients, both in the ACHD (82.3%) and in pediatric centers (89%), were seen annually and had a physical examination, electrocardiogram, and echocardiogram performed at each visit. However, unsatisfactory utilization of tests identifying the early stages of Fontan circulation failure (cardiopulmonary exercise tests, cardiac magnetic resonance, liver biochemistry and imaging, detection of protein-losing enteropathy) was observed.

**Conclusions:** The results of the study showed that there is no unified surveillance approach for Fontan patients in Poland. The practice of care for adults differs from that of adolescents.

**Key words:** adult congenital heart diseases, Fontan surgery, practice of care, univentricular heart

## INTRODUCTION

For the last decades, advances in surgical and medical care have led to rapid growth in the population of adults living with even complex congenital heart disease (adult congenital heart disease, ACHD) [1, 2]. Among them, the most challenging for both the healthcare system and professionals and potentially the most vulnerable to deficiency of care are those who underwent palliative procedures that eventually transformed to the Fontan circulation [3]. After the procedure, the systemic veins directly enter the pulmonary arteries, leading to an improvement in arterial oxygen saturation and a reduction of systemic ventricular volume overload. To maintain its function, the Fontan circulation increases systemic venous pressure, resulting in gradual elevation of pulmonary vascular resistance and decreased cardiac output due to complex pathophysiological mechanisms. Eventually, they cause progressive cardiac dysfunction and deterioration of pulmonary, liver, and renal structure and function. However, many pathophysiological relationships have not been fully elucidated [3–5].

Multiorgan damage that potentially may develop requires meticulous medical care for Fontan patients, including regular surveillance tests not only dedicated to cardiovascular status, but also for already identified Fontan-specific complications and comorbidities such as plastic bronchitis, liver dysfunction, protein losing enteropathy (PLE), kidney disease, neurodevelopmental delay and others (Table 1) [3–5].

Current recommendations for the management of patients after Fontan operation are summarized in the ESC guidelines and AHA/ACC scientific statements [1–3]. Experts from both European and American countries emphasize the need for serial, highly qualified care for these patients for the reasons mentioned above. ESC guidelines recommend careful evaluation of Fontan patients in specialized centers with predefined requirements for multispecialized personnel, diagnostic and therapeutic modalities [1]. The AHA/ACC statement recommends cardiovascular evaluation at set intervals in all age groups of Fontan patients. Its frequency should be adapted to the age of the patient with recommendations of one to two years in adulthood. The necessity of end-organ system surveillance testing is also emphasized [3].

As in Poland, there is no organized, coordinated health care system dedicated to ACHD, including patients after Fontan surgery, our objective was to evaluate current practice of care for Fontan patients in our country based on a multicenter survey. We conducted an observational study in which we investigated the frequency and selection of surveillance tests in large centers specialized in ACHD in Poland and compared them with international recommendations. Furthermore, data from ACHD centers were contrasted with those of pediatric centers that care for adolescents between 14 and 18 years of age (AdolCHD centers).

## METHODS

The study was carried out by the Polish Cardiac Society Working Group on Congenital Heart Diseases with the support of the Polish Cardiac Society Clinical Initiatives Committee and the Polish Cardiac Society. The center of the principal investigator who coordinated the study was the Department of Congenital Heart Diseases of the National Institute of Cardiology, Warszawa, Poland.

Eight centers for adolescent and adult Fontan patients were invited to participate in the survey and six accepted the invitation. Among them, three centers serve only adults, one was a purely pediatric center, while two cared for both adults and adolescents. For the purpose of the study, the latter two were divided into two separate centers which enrolled adults and adolescents separately. Eventually, we included eight centers in the final analysis: five ACHD centers and three AdolCHD centers (Figure 1).

After signing the agreement and obtaining the consent of the relevant ethics committees, access to an anonymous web-based survey was sent to the authorized cardiologists of each participating center.

The first center (the center of the principal investigator) was opened on the platform in September 2021. Access to the survey was closed in October 2022. It consisted of basic demographic data of the patient, patient care (regular or irregular), as well as type and frequency of surveillance tests in each center. The survey was created based on current ESC and AHA guidelines. The proposed surveillance tests included evaluation of the cardiovascular, respiratory, lymphatic, and nervous systems, as well as kidney and liver function. The time intervals used initially to assess the frequency of the test in the survey (<12 months, every 12 months, 12–18 months, 18–24 months and > 24 months) were later grouped into three categories to make the data obtained more comparable both with the current guidelines and with the analyzes already conducted in the ACHD or specifically the Fontan population in different regions [1–5]. Therefore, for the purpose of the study, the time intervals to evaluate the frequency of the test were the following: up to 18 months (consistent with the time intervals: <12 months, 12 months and 12–18 months); every two years (consistent with the time intervals 18-24 months) and more than two years (consistent with the time interval >24 months).

To estimate the quality of patient care and interpret the results, we developed the Fontan Surveillance Score (FSS). The model includes 19 questions about the type and frequency of surveillance tests (Table 2). In the FSS, the time intervals of patient surveillance were defined according to the current ESC and AHA guidelines.

## Statistical analysis

Data analysis was carried out using the statistical software IBM SPSS Statistics 23 (IBM Corp. Released 2015. IBM SPSS Statistics for Windows, version 23.0. Armonk, NY: IBM Corp.). Data are expressed as mean (standard deviation [SD]) or median (interquartile range [IQR]), unless otherwise noted. The Kolmogorov-Smirnov test was used to assess the conformity with a normal distribution. To compare the distribution of numerical variables between the groups, the Mann-Whitney test, or Kruskal Wallis test were used. The  $\chi^2$  test was used for categorical data. A two-sided  $P < 0.05$  was considered statistically significant.

## RESULTS

The survey was completed by 8 centers. There was a considerable difference in the number of Fontan patients cared for in each center (Figure 1).

A total of 398 patients after the Fontan operation were included in the study. The group consisted of 243 adults (age >18 years) and 155 adolescents (age 14–18 years) (Figure 1). The characteristics of the patients are presented in Table 3.

ACHD and AdolCHD centers did not differ in terms of the percentage of patients under regular surveillance, however, adolescents were seen more often. Most of the patients were under routine surveillance, both in the hospital and in the outpatient clinic. The practice of care of adult patients differed from that of adolescents. Adolescent patients were seen more often in the outpatient clinic, whereas adults patients were hospitalized more frequently (Table 4).

The median score for all centers was 13 points (interquartile range: 7–14 points). There was a difference in FSS between the ACHD and AdolCHD centers ( $P < 0.001$ ) (Table 3, Figure 2A). Subsequently, we compared all centers in terms of the FSS achieved and found a difference in the median FSS between centers ( $P = 0.005$ ) (Figure 2B).

The choice and frequency of the tests according to FSS in all patients, as well as a comparison between the ACHD and AdolCHD centers, is presented in Figure 3.

Most of patients, both in the ACHD and AdolCHD centers, have been seen annually during a routine medical visit (82.3% and 89%;  $P = 0.46$ , respectively) and have had a physical examination, electrocardiogram, and echocardiogram performed at each annual visit (Figure 3).

Holter monitoring was performed annually in 57.5% of the patients, more frequently in adolescents (51.4% vs. 61.7%;  $P = 0.002$ ) (Figure 3).

Blood tests were performed annually in 67.6% of the patients, more frequently in adolescent patients than in adults (Figure 3). In all patients, both at the ACHD and AdolCHD centers, the level of NT-proBNP was evaluated annually.

The annual cardiopulmonary exercise test (CPET) was performed in 44% of the patients with no differences between the compared groups (45.7% vs. 41.3%;  $P = 0.39$ ) (Figure 3).

Cardiac magnetic resonance (CMR) was performed every two years in 5.3% of the patients, more often in adult patients than in adolescents (7.8% vs. 1.3 %;  $P = 0.004$ ) (Figure 3).

The percentage of patients who underwent routine annual liver imaging was low in both the ACHD and AdolCHD centers (6.6% vs. 7.8%;  $P = 0.68$ , respectively) (Figure 3).

PLE detection was performed annually in 52.8% of the patients,—more frequently in adolescents than in adults (77.4% vs. 31.7%;  $P < 0.001$ ) (Figure 3). It should be noted that in adult Fontan patients, the level of fecal alpha-1 antitrypsin was measured in these patients who were under the care of centers serving both adolescents and adults.

In only 18.8% of the patients, chest radiographs were performed routinely, more frequently in adolescents than in adults (26.5% vs. 14%;  $P = 0.002$ ) (Figure 3).

The availability of invasive procedures (cardiac catheterization, ablation, cardiac surgery) and the possibility of emergency admission, in the case of clinical indications, was high in all centers (Figure 3).

## DISCUSSION

Improvements in the treatment strategy of pediatric Fontan patients have reduced their mortality to the point where a large number of patients survive to adulthood [2, 3]. Studies from Australia and Canada have shown the positive impact of specialized supraregional tertiary centers on adherence as a measure of process quality and overall mortality in patients with ACHD. As a consequence, consolidation of ACHD care and the creation of relevant centers have been implemented in several countries [6–8]. According to the ESC guidelines, one of these centers should serve the general population [1].

In Poland, there is no mandatory well-organized care system dedicated to ACHD. This may limit access to qualified health care, particularly for the most demanding and heterogeneous group, such as those who underwent Fontan palliation. Therefore, we investigate to what extent the current service offers sufficient care for these patients in Poland.

To objectively compare centers, we have developed a model that assesses the quality of patient care, FSS. The higher score is consistent with better care, with a maximum of 19 points. Centers that provided continuous care from the pediatric period until 18 years of age achieved a higher

FSS compared to ACHD centers. The most likely it resulted from a greater experience and familiarity with complex post-Fontan problems among pediatric cardiologists. However, when comparing the FSS between all centers and between ACHD and AdolCHD centers, we discovered a high variability in the score between all sites. It should be noted that centers that cared for adults and adolescents achieved a higher score than ACHD centers, highlighting the need for organized care in the transition period and sustained cooperation between pediatric cardiologists and those who care for adults. Although the practice for care of adolescent Fontan patients differed from that of adults patients in many aspects, we observed a similarly high percentage of annual routine visits with physical examination, electrocardiogram, and echocardiogram. Adolescents were seen more often in periods of less than 12 months, while adult patients were seen in longer intervals. There were also more outpatient visits in adolescent group as compared to adults who were hospitalized more frequently. Taking into account the fact that adolescents had higher FSS, the model of care proposed for patients with ACHD appears to be more expensive and less effective.

The survey results also showed a substantial difference in the diagnostic approach between cardiologists caring for adolescent and adult patients. In the latter case, oxygen saturation, 12 lead electrocardiogram, Holter ECG, chest radiograph, and cardiac catheterization were performed less frequently in adults. Although the availability of invasive procedures in pediatric patients is quite satisfactory, adult patients have less access to invasive diagnostics and percutaneous procedures. Importantly, cardiac surgery dedicated to ACHD was not available in all centers.

The role of CPET in ACHD surveillance is well established. Since exertional performance deteriorates during follow-up in Fontan patients, CPET is extremely useful for baseline and serial functional evaluation. The deterioration in the CPET parameters observed during follow-up helps eliminate patient symptoms and optimize decisions about cardiac catheterization and/or electrophysiological interrogation. The CPET examination plays an important role in planning pregnancy (if considered and not contraindicated) and physical activity [9,10]. The American guidelines recommend performing CPET every 1 to 2 years in adults and every 1 to 3 years in adolescents. In our study, less than 50% of the patients had performed CPET annually in both groups [3].

**CMR** offers a detailed and accurate non-invasive method of evaluating cardiac anatomy and function that is often altered after interventions, particularly in patients with complex CHD [11]. The American guidelines recommend performing CMR every 2–3 years in Fontan patients [3]. Our study found an extremely low percentage of patients in whom CMR was



performed every 2 years (5.1% of patients) and adults had CMR performed more often than adolescents (Figure 3).

The hemodynamics of the liver is highly affected by elevated venous pressure, which is a consequence of Fontan circulation. In this population, congestive hepatopathy, fibrosis cirrhosis, and hepatocellular carcinoma (with a prevalence between 1.5% and 5% of patients) are reported, which are associated with increased morbidity and mortality. Therefore, regular abdominal ultrasound focused on liver evaluation, performed at least 1–2 years is crucial in the routine evaluation of Fontan patients and may reduce patient mortality [12, 13]. Regarding liver morphology and function, the survey revealed a low number of relevant tests performed in both groups, with a significantly higher percentage of patients having annual evaluation in the adolescent group (Figure 3).

Another particularly perplexing and enigmatic problem with significant morbidity and mortality is PLE. The disease can strike at any time, from weeks to years after Fontan surgery, and its pathophysiology is unknown. PLE is defined as an abnormal loss of serum proteins in the lumen of the gastrointestinal tract that hosts serious clinical manifestations and gastrointestinal symptoms. The gold standard test for the diagnosis of PLE is an abnormal clearance of alpha-1-antitrypsin from the stool [14, 15]. The American experts found it reasonable to consider surveillance tests every 1 to 3 years in Fontan and every 1 to 2 years in adults with Fontan circulation [3]. In our study, the percentage of patients who are regularly tested for PLE was very low in the adult group (31.7%), while much more satisfactory in adolescents (77.4%).

Therefore, our findings confirm the discrepancy between pediatric care for Fontan adolescents and adult Fontan patients. This indicates an urgent need to establish a relevant number of specialized ACHD centers in Poland according to the ESC recommendations that could provide complex professional care to Fontan patients, including a transition period of care served by a team of pediatric and adult cardiologists. It is well established that poor health care transition has a significant impact on the prognosis and well-being of ACHD patients. A systematic, multifaceted, collaborative approach to transition is critical to improving lifelong care and providing optimal quality of life and prognosis of Fontan patients [1, 3, 16].

In 2019 di Maria et al. presented data from the multiinstitutional survey assessing the surveillance testing and preventive care after Fontan operation in the US. The survey was carried out in 11 centers specializing in CHD and covered the population of 1 500–2 000 Fontan patients [17].

Similarly to our study, they found considerable variability in the surveillance testing regimen and management strategy after Fontan operation in different national specialized centers. When comparing the findings of our study and those of the US study, it turned out that Polish post-Fontan patients are followed up more frequently, with more frequent detailed assessment of the cardiovascular system (echocardiogram, Holter ECG, CPET, NT-pro-BNP), with the exception of CMR, which was performed more frequently in American patients [17]. However, in Poland, screening for Fontan circulation complications, such as blood tests, liver assessment, or PLE diagnostics, were performed less frequently. It should be noted that, unlike Poland, all American centers that care for patients with ACHD have access to percutaneous and surgical procedures. The comparison of the discussed studies results is presented in [Table 5](#). The data of Polish and American multiinstitutional surveys show clearly that the considerable variability in the surveillance testing regimen and lack of standardized management strategy after Fontan procedure is a worldwide problem. Therefore, there is an urgent need for precise surveillance guidelines, defining quality metrics, establishing collaborative practice, and conducting prospective research to allow health professionals to take care better and more effectively after complex Fontan patients. As shown by Di Maria et al. [18]. The dedicated Fontan multidisciplinary clinic with many available subspecialists improves adherence to the therapy and the quality of treatment recommended by the guidelines.

There are minor limitations of the study. The findings are mainly interesting for Polish cardiologists and Polish healthcare authorities. However, it is noteworthy that our data along with the results of the American survey discover the considerable variability in the surveillance testing regimen and the lack of standardized management strategy after the Fontan procedure as a worldwide problem and stimulate the need for discussion, also on the international level. Furthermore, the introduction of FSS seems to be a valuable and simple approach that provides a way of comparison between various countries in order to standardize it and outcomes optimization.

Second, in our study we did not take into account the clinical condition of Fontan patients. As a result of variable primary patient characteristics, the clinical status of Fontan patients can be different for each patient what influences the diagnostic approach. As presented in our previous studies, sophisticated imaging techniques provide new data on patients' clinical status and prognosis, therefore influencing the sequence of further assessment [19, 20]. Additionally, the sequelae of the procedures may differ depending on the age of the patients. These variables may affect the final results of our survey. However, the purpose of the study was not a qualitative assessment of the care, but a definition of current practice. We hope that our findings

and conclusions may start the discussion about need for the implementation of coordinated care dedicated to Fontan patients in Poland, as it was successfully organized for patients after acute coronary syndrome [21].

In summary, for the first time, the practice of care of patients after the Fontan operation in Poland was systematically evaluated. As variability in the surveillance testing regimen was shown in all participating centers, it was shown that there is no systemic surveillance approach for Fontan patients. The practice of care for Fontan patients differed from that of adults in many aspects, including the frequency of routine visits and the proposed diagnostic evaluation.

In general, the frequency of routine visits and the basic assessment of cardiovascular status were consistent with current guidelines in all centers, while the use of diagnostic tests for Fontan-associated complications and comorbidities was unsatisfactory. Patients with ACHD also appeared to have a worse availability of cardiac surgery and invasive procedures, which are a crucial part of treatment in those patients also in adulthood.

The findings of our study clearly defined these aspects of Fontan patients' care that need urgent improvement such as an increase in the frequency of regularly performed CPET, CMR, abdominal ultrasound, screening for PLE what should be introduced as a priority. Our findings emphasize a need for discussion among experts and national authorities to introduce systemic solutions for the organization of the adult transition period, the management strategy, and the education that would meet the requirements of the current guidelines and provide the best possible care for patients after Fontan surgery.

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**Table 1.** Fontan circulation complications and comorbidities

Heart and circulatory system	
<ul style="list-style-type: none"> <li>• Heart and circulatory failure</li> <li>• Worsening of ventricular function</li> <li>• Cyanosis</li> <li>• Atrioventricular valve regurgitation</li> <li>• Arrhythmias</li> </ul>	
Venous and lymphatic congestion	
<ul style="list-style-type: none"> <li>• Protein-losing enteropathy</li> <li>• Plastic bronchitis</li> </ul>	
Fontan-associated liver disease	
<ul style="list-style-type: none"> <li>• Liver fibrosis</li> <li>• Cirrhosis</li> <li>• Hepatocellular carcinoma</li> </ul>	
Others	
<ul style="list-style-type: none"> <li>• Kidney disease</li> <li>• Abnormalities in body composition, bone structure, and growth</li> <li>• Cognitive, neuropsychological, and behavioral deficits</li> </ul>	

**Table 2. Fontan Surveillance Score**

Fontan Surveillance Score (FSS)	Yes = 1 point; No = 0 points
Routine visits every year	Yes/No
Cardiopulmonary testing	
Physical Examination including oxygen saturation	Yes/No
Electrocardiogram every year	Yes/No
Echocardiogram every year	Yes/No
Holter ECG every year	Yes/No
Cardiopulmonary exercise test every year	Yes/No
Chest radiograph every year	Yes/No
Cardiac magnetic resonance imaging every 2 years	Yes/No
Cardiac catheterization in the case of clinical indication	Yes/No

Hepatic imaging	
Ultrasound, magnetic resonance, or computed tomography every year	Yes/No
Serum studies	
Complete blood count every year	Yes/No
Cholesterol level every year	Yes/No
Creatinine level every year	Yes/No
Liver function test every year	Yes/No
NT-proBNP every year	Yes/No
Screening for protein losing enteropathy	Yes/No
Availability of emergency admission	Yes/No
Availability of invasive procedures if indicated	
Electrophysiology (ablation, etc.).	Yes/No
Cardiac surgery	Yes/No
<b>Overall score (max. 19 points)</b>	

**Table 3.** Patients' characteristics

	All centers	AChD centers	AdolCHD centers
No. of patients	398	243	155
Sex, female/male	176/222	108/135	68/87
Mean age, years	22.5 (7.9)	26.8 (7.4)	15.9 (1.1)
Age range, years, median (IQR)	14–54	18–54	14–18

Data are presented as mean and standard deviation

**Table 4.** Routine medical visits in patients after the Fontan operation.

	All centers	AChD centers	AdolCHD centers	<i>P</i> -value
No. of patients	398	243	155	
Routine visits, n (%)	373 (93.7)	226 (92.5)	147 (94.8)	0.46
Hospitalization and outpatient clinic, n (%)	302 (81)	174 (77)	128 (87.1)	0.015
Hospitalization only, n	42 (11.3)	39 (17.2)	3 (2.0)	<0.001



(%)				
Outpatient clinic only, n (%)	29 (7.7)	13 (5.8)	16 (10.9)	0.070
Frequency				
Up to 18 months, n (%)	338 (90.6)	200 (88.6)	138 (93,9)	0.082
18–24 moths, n (%)	20 (5.4)	15 (6.6)	5 (3.2)	0.18
>24 months, n (%)	15 (4.0)	11 (4.8)	4 (2.6)	0.30

**Table 5.** Comparison of the Polish study with the results of the REDCap survey, Di Maria et al. [17]

Every <18 months	Current study	REDCap Survey, US [17]
Number of participating centers	8	11
Routine visits	84.9%	73%
Electrocardiogram	85.9%	90.9%
Echocardiogram	79.1%	72.7%
Holter ECG	57.5%	27.3%
Cardiopulmonary exercise test	44%	9.1%
NT-proBNP	100%	54.5%
Liver biochemistry	57.8%	72.3%
Liver imaging	7.0%	9.1%
Creatinine level	66.6%	72.3%
Complete blood count	7.3%	72.3%
Cholesterol level	50.5%	72.3%
Alpha-1-antitripsin	63.8%	72.3%
Chest radiograph	18.8%	0%
Cardiac magnetic resonance every 2 years	5.3%	9.1%
Availability of cardiac catheterization	366 (92%)	398 (100%)
Availability of cardiac ablation	398 (100%)	398 (100%)
Availability of cardiac surgery	366 (92%)	398 (100%)
Availability of urgent	398 (100%)	398 (100%)

hospitalization

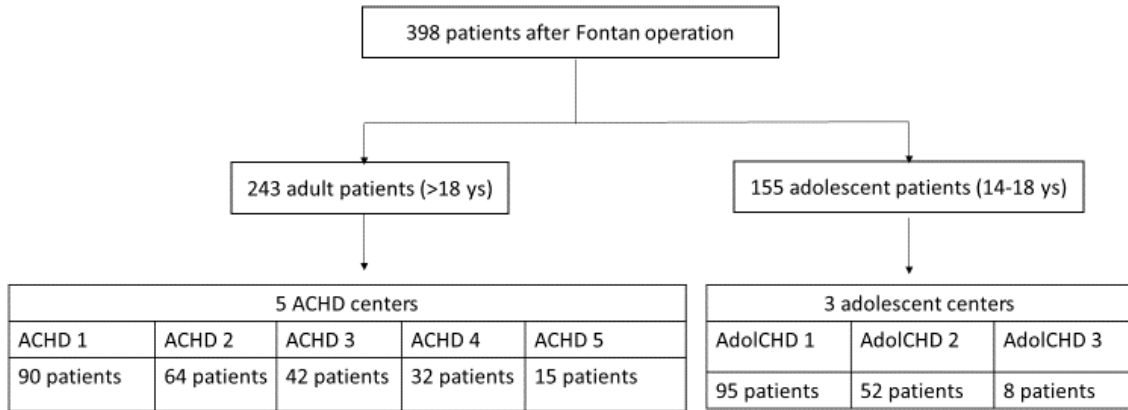
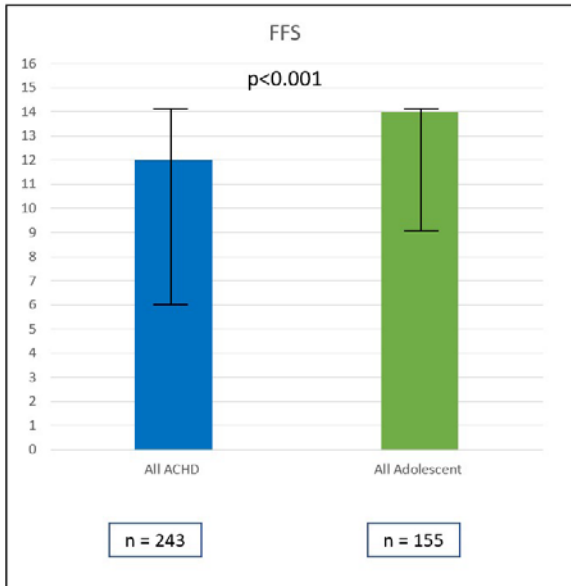


Figure 1

a. Comparison of FFS between ACHD and AdolACHD centers



b. Comparison of FFS among participating centers

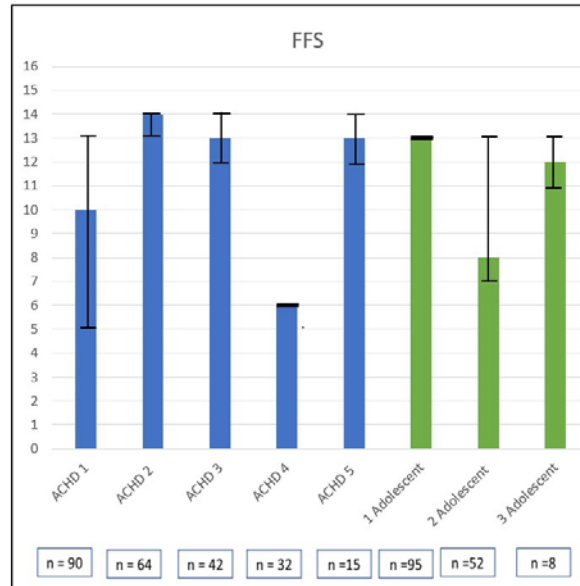


Figure 2

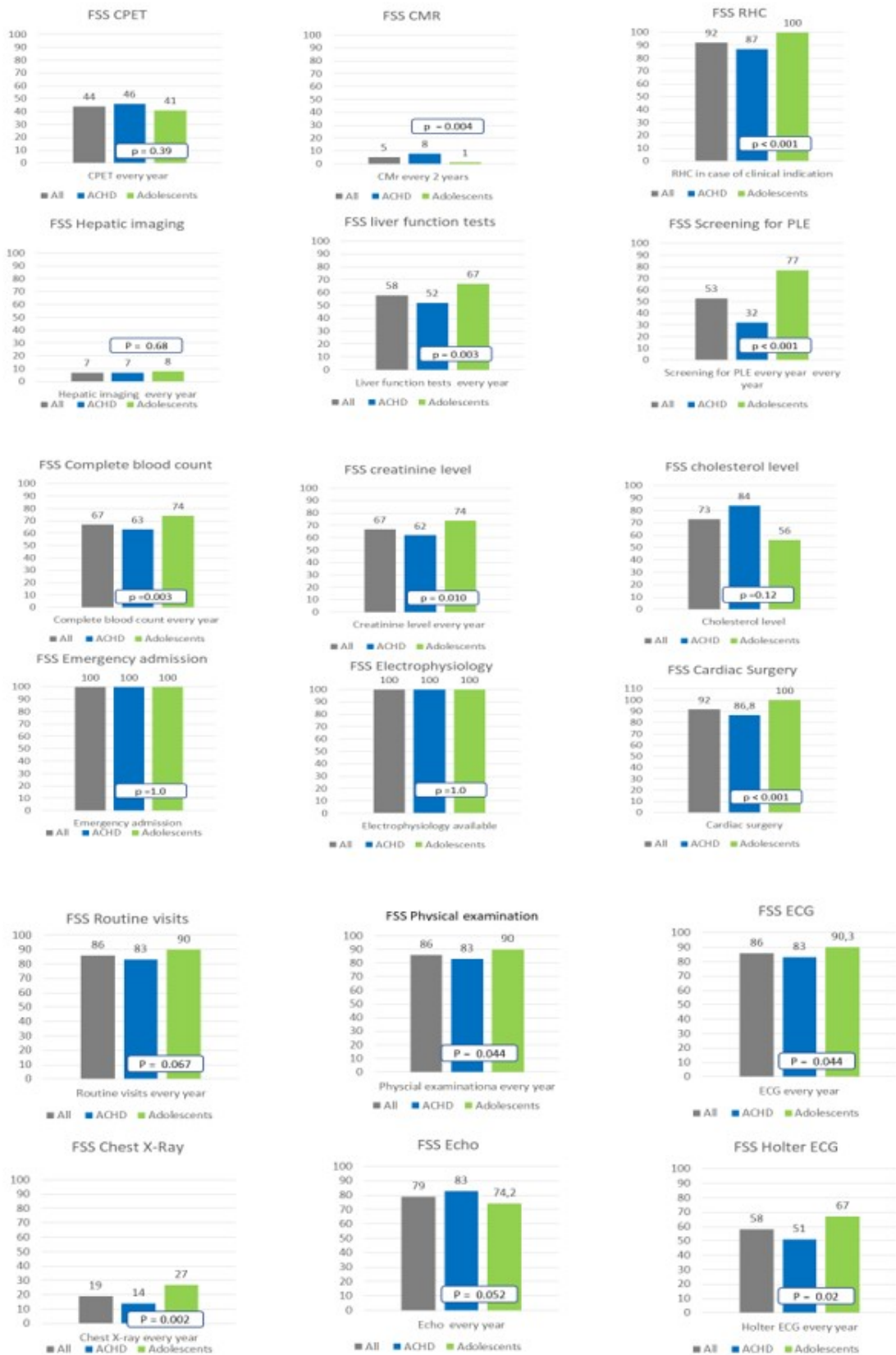


Figure 3