

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

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ABSTRACT

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

Aims: Our study aimed to evaluate the current practice of care for Fontan patients in Poland using a multicenter survey.

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

Results: We included in the study 398 Fontan patients (243 adults and 155 adolescents [aged 14–18 years]). The median FSS was 13 points with variability between 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 pediatric centers (89%), were seen annually and had a physical examination, electrocardiogram, and echocardiogram performed at each visit. However, we observed unsatisfactory utilization of tests identifying early stages of Fontan circulation failure (cardiopulmonary exercise tests, cardiac magnetic resonance, liver biochemistry and imaging, detection of protein-losing enteropathy).

Conclusions: Our results 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

In 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, patients who underwent palliative proce-

dures that eventually transformed into the Fontan circulation are the most challenging for both the healthcare system and professionals. These patients are also potentially the most vulnerable to shortage of care [3]. After the procedure, the systemic veins enter directly the pulmonary arteries, leading to an

WHAT'S NEW?

For the first time, the practice of care for patients after the Fontan operation in Poland was systematically evaluated. In general, the frequency of routine visits and basic cardiovascular status assessments 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 early stages of associated complications are rarely used. The practice of 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, our study demonstrated 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.

Table 1. Fontan circulation complications and comorbidities

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

improvement in arterial oxygen saturation and a reduction in 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 in Fontan patients requires meticulous medical care, including regular surveillance tests not only dedicated to cardiovascular status but also 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 the Fontan operation are summarized in the European Society of Cardiology (ESC) guidelines and American Heart Association/American College of Cardiology (AHA/ACC) scientific statements [1–3]. Experts from both Europe and America emphasize the need for serial highly qualified care for these patients for the reasons mentioned above. The ESC guidelines recommend careful evaluation

of Fontan patients in specialized centers with predefined requirements for multi-specialized personnel, and 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 patient's age, with a recommendation of one visit every year or every two years in adulthood. The necessity for end-organ system surveillance testing is also emphasized [3].

As there is no organized, coordinated healthcare system in Poland dedicated to ACHD and including patients after Fontan surgery, our objective was to evaluate the 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 Working Group on Congenital Heart Diseases of the Polish Cardiac Society with the support of the Clinical Initiatives Committee of 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. In this 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 an agreement and obtaining approval from the relevant ethics committees, access to an anonymous web-based survey was sent to the authorized cardiologists of each participating center.

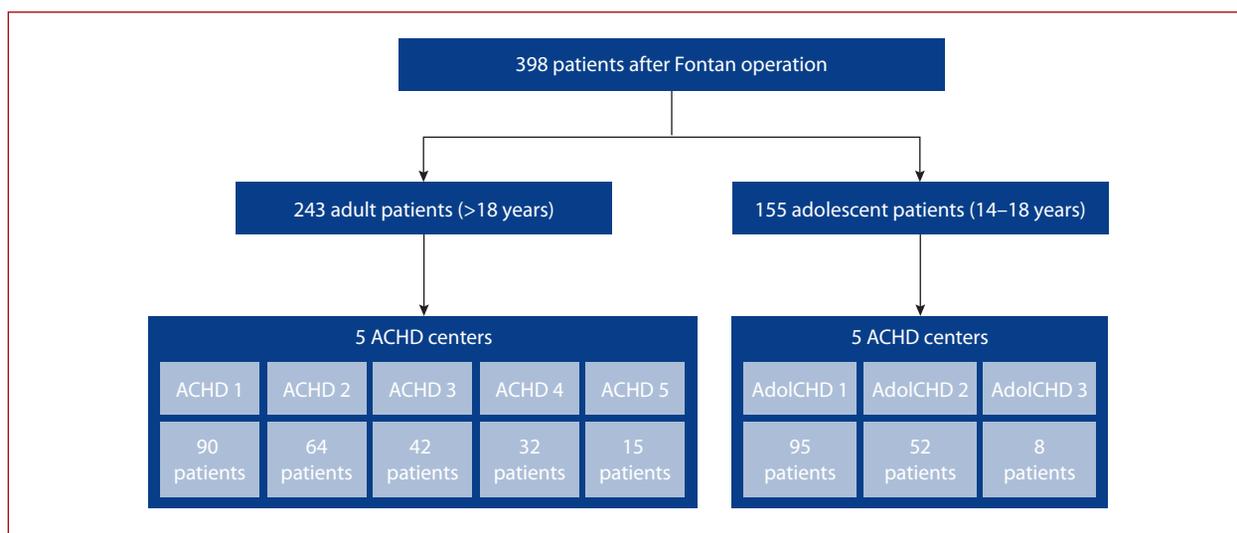


Figure 1. Study flowchart. Number of patients from participating centers

Abbreviations: ACHD, adult congenital heart disease; AdoIACHD, pediatric centers that care for adolescents between 14 and 18 years of age

The survey was opened on the platform from September 2021 till October 2022. The survey consisted of questions about basic demographic patient data, patient care (regular or irregular), as well as type and frequency of surveillance tests in each center. The survey was based on the 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 with both the current guidelines and studies already conducted in the ACHD population or, specifically, the Fontan population in various regions [1–5]. Therefore, in our study, we used the following time intervals to evaluate the frequency of the tests: 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 were expressed as means (standard deviation [SD]) or medians (interquartile range [IQR]) unless otherwise

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 cell 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
Overall score (max. 19 points)	

Abbreviations: NT-proBNP, N-terminal pro-B-type natriuretic peptide

noted. The Kolmogorov-Smirnov test was used to assess the conformity with normal distribution. To compare the distribution of numerical variables between groups, the Mann-Whitney or Kruskal-Wallis tests were used. The χ^2 test was used for categorical data. A two-sided $P < 0.05$ was considered statistically significant.

Table 3. Patient characteristics

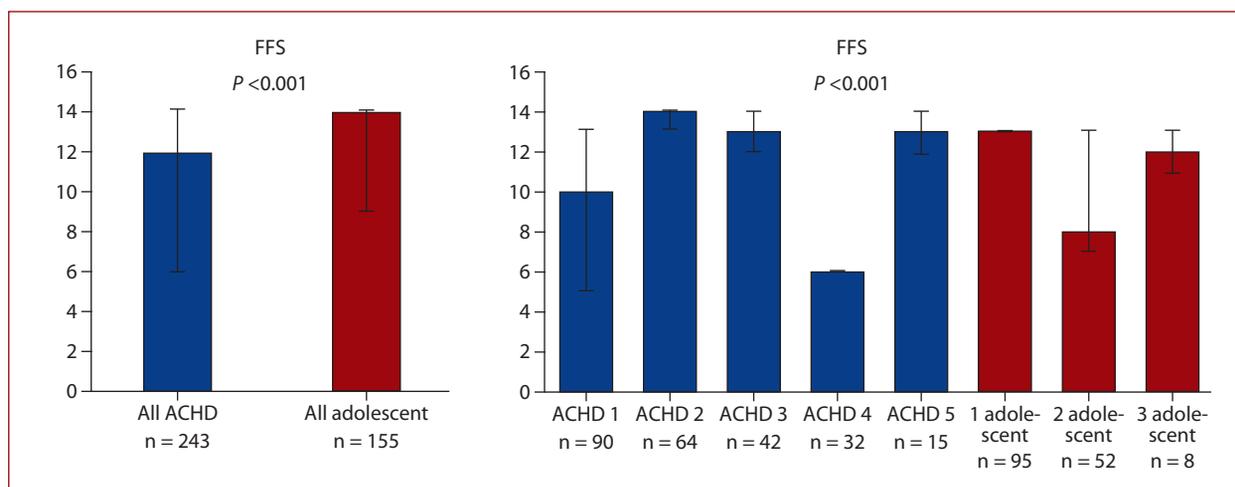
	All centers	ACHD centers	AdolCHD centers
No. of patients	398	243	155
Sex, female/male	176/222	108/135	68/87
Mean age, years, mean (SD)	22.5 (7.9)	26.8 (7.4)	15.9 (1.1)
Age range, years	14–54	18–54	14–18

Abbreviations: SD, standard deviation; other — see Figure 1

Table 4. Routine medical visits of patients after the Fontan operation

	All centers	ACHD centers	AdolCHD centers	P-value
No. of patients	398	243	155	
Routine visits, n (%)	373 (93.7)	226 (92.5)	147 (94.8)	0.46
Hospitalization and outpatient center, 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 center 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 months, 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

Abbreviations — see Figure 1

**Figure 2.** Comparison of the Fontan Surveillance Score (FSS) in the participating centers

Abbreviations: see Figure 1

RESULTS

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

A total of 398 patients after the Fontan operation were included in the study. The group included 243 adults (age >18 years) and 155 adolescents (age 14–18 years) (Figure 1). The patient characteristics 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 center. The practice of care for adult patients differed from that for adolescents. Adolescent patients were seen more often in the outpatient center, whereas adult 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 the 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 the FSS, in all patients, as well as a comparison between the ACHD and AdolCHD centers, are presented in Figure 3.

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

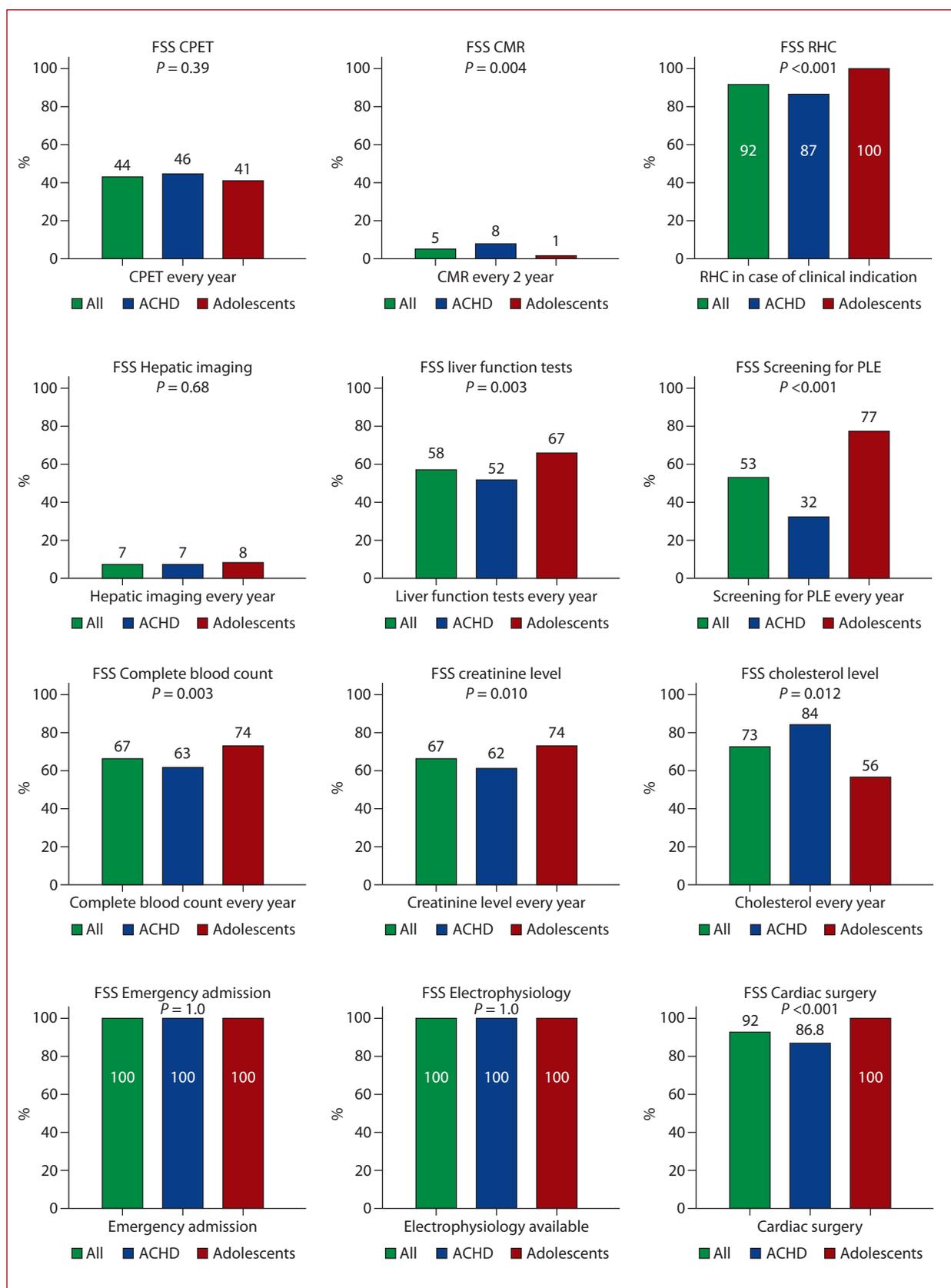


Figure 3. Choice and frequency of testing according to the Fontan Surveillance Score (FSS) in all patients and comparison between ACHD and AdolCHD centers

Abbreviations: CMR, cardiac magnetic resonance; CPET, cardiopulmonary exercise test; ECHO, echocardiography; ECG, electrocardiography; PLE, protein-losing enteropathy; RHC, right heart catheterization; other — see Figure 1

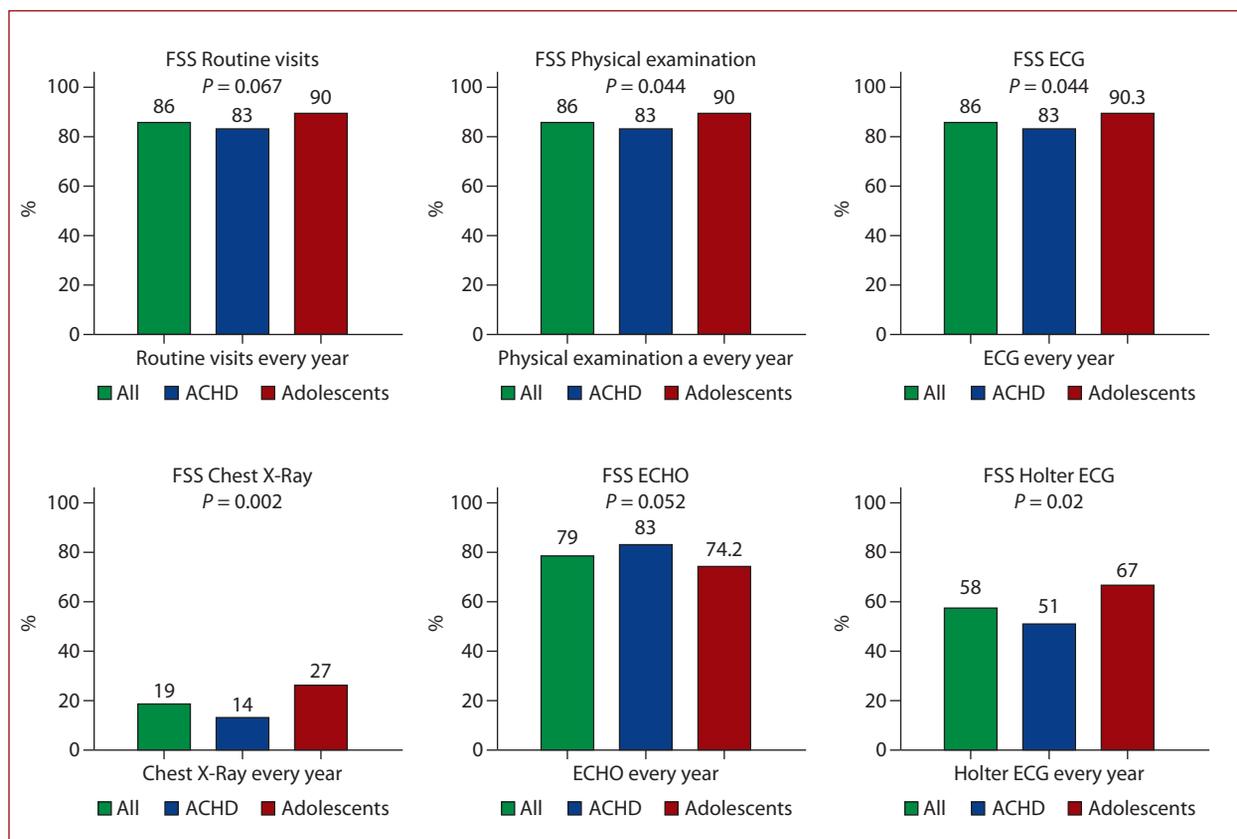


Figure 3 (cont.). Choice and frequency of testing according to the Fontan Surveillance Score (FSS) in all patients and comparison between ACHD and AdolCHD centers

Abbreviations: CMR, cardiac magnetic resonance; CPET, cardiopulmonary exercise test; ECHO, echocardiography; ECG, electrocardiography; PLE, protein-losing enteropathy; RHC, right heart catheterization; other — see [Figure 1](#)

Holter monitoring was performed annually in 57.5% of the patients, and 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 in the ACHD and AdolCHD centers, the NT-proBNP level 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%, respectively; $P = 0.68$) ([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 anti-trypsin 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 capacity for 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 ACHD patients. As a consequence, several countries consolidated ACHD care and created relevant centers [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 healthcare, particularly for the most demanding and heterogeneous group, such as patients who underwent Fontan palliation. Therefore, we investigated to what extent the current service offers sufficient care for these patients.

To objectively compare centers, we have developed a model that assesses the quality of patient care, the FSS. The higher score is consistent with better care, with a maximum of 19 points. The centers that provided continuous care from the pediatric period until 18 years of age achieved a higher FSS compared to the ACHD centers. Most likely the difference 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 scores 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 for adult patients in many aspects, we observed a similar 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 at longer intervals. There were also more outpatient visits in the adolescent group as compared to adults who were hospitalized more frequently. Taking into account the fact that adolescent centers had a higher FSS, the model of care proposed for ACHD patients 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, in adults, oxygen saturation, 12-lead electrocardiogram, Holter ECG, chest radiograph, and cardiac catheterization were performed less frequently. 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 the CPET in ACHD surveillance is well established. Since exertional performance deteriorates during follow-up in Fontan patients, the CPET is extremely useful for baseline and serial functional evaluation. The deterioration in CPET parameters observed during follow-up helps eliminate patient symptoms and optimize decisions about cardiac catheterization and/or electrophysiological interrogation. CPET examination plays an important role in planning pregnancy (if considered and not contraindicated) and physical activity [9,10]. The American guidelines recommend performing the CPET every 1 to 2 years in adults and every 1 to 3 years in adolescents. In our study, fewer than 50% of the patients in both groups had performed the CPET annually [3].

CMR offers a detailed and accurate non-invasive method of evaluating cardiac anatomy and function, which are often altered after interventions, particularly in patients with complex CHD [11]. The American guidelines recom-

mend 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 are 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 every 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 showed 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 triggers serious clinical manifestations and gastrointestinal symptoms. The gold standard test for the diagnosis of PLE is abnormal alpha-1-antitrypsin clearance in the stool [14, 15]. American experts found it reasonable to consider surveillance tests every 1 to 3 years in Fontan patients 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 results were obtained in adolescents (77.4%).

Therefore, our findings confirm a discrepancy between pediatric care for Fontan adolescents and adult Fontan patients. This indicates an urgent need to establish a proper 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 that should be overseen by a team of pediatric and adult cardiologists. It is well established that poor healthcare transition has a significant impact on prognosis and well-being of ACHD patients. A systematic, multifaceted, collaborative approach to transition is critical in improving lifelong care and providing optimal quality of life and prognosis for Fontan patients [1, 3, 16].

In 2019 Di Maria et al. [17] presented data from a multi-institutional survey assessing the surveillance testing and preventive care after the 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. Similar to our study, the authors found considerable variability in the surveillance testing regimen and management strategy after Fontan operation in different national specialized centers. When comparing the findings

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-antitrypsin	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 hospitalization	398 (100%)	398 (100%)

Abbreviations: see [Figures 2 and 3](#)

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-proBNP), except for 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, was 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 from the Polish and American multi-institutional surveys show clearly that the considerable variability in the surveillance testing regimen and lack of standardized management strategy after the 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 better and more effective care for complex Fontan patients. As shown by Di Maria et al. [18], a dedicated Fontan center with many multidisciplinary subspecialists improves adherence to the therapy and the quality of treatment recommended by the guidelines.

There are minor limitations of this study. Our 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, shows considerable variability in the surveillance testing regimen and the lack of standardized management strategy after the Fontan procedure as a worldwide problem. This also indicates the need for discussion on the international level. Furthermore, the introduction of the FSS seems to be a sim-

ple and valuable approach for standardized comparison between various countries to optimize outcomes.

Second, in our study, we did not take into account the clinical condition of Fontan patients. As a result of variable primary patient characteristics, clinical status of Fontan patients can be different for each patient, which 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 patient's age. These variables may have affected the final results of our survey. However, the purpose of the study was not a qualitative care assessment but defining the current practice. We hope that our findings and conclusions may start a discussion about the need for implementing in Poland coordinated care dedicated to Fontan patients similar to that successfully organized for patients after acute coronary syndrome [21].

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

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

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

Article information

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