The need for a unified approach to the univentricular circulation: Current practice of care for adolescent and adult patients after Fontan surgery in Poland

Paul Clift[1, 2], Lucy Cowie[1], Sasha Douglas[1, 2]

[1]Adult Congenital Heart Unit, Department of Cardiology, University Hospital Birmingham, Birmingham, United Kingdom
[2]Institute of Cardiovascular Sciences, University of Birmingham, Birmingham, United Kingdom

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Correspondence to:
Paul Clift, MBBS, MD, FRCP, Adult Congenital Heart Unit, Department of Cardiology, University Hospital Birmingham, B15 2TH Birmingham, United Kingdom
phone: +44 12 137 247 31,
e-mail: pclift@nhs.net

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INTRODUCTION

The Fontan operation is a palliative procedure carried out in univentricular congenital heart disease where biventricular repair is not possible. The concept involves directing systemic blood flow to the pulmonary circulation without active ventricular propulsion. The surgery has evolved since Francis Fontan described his initial series [1], and this has led to increasing survivorship through adolescence to adulthood. Whilst life-prolonging, Fontan surgery has consequences leading to late cardiac and extra-cardiac morbidity, which physicians caring for these patients need to detect and manage.

COMPLICATIONS OF A FONTAN CIRCULATION

An effective Fontan relies on low pulmonary vascular resistance (PVR) and elevated systemic venous pressure to drive transpulmonary blood flow. Systemic venous hypertension, however, can lead to direct effects on the liver, with a degree of fibrosis seen in all Fontan survivors [2] and abnormalities of the peripheral venous and lymphatic circulation. However, whichever organ is studied, there is evidence of an impact due to the effect of the Fontan circulation and the multiple surgeries performed in early life (Table 1). In our experience, educational outcomes have been largely good, and adult employment is typical, however, there is a risk of long-term learning deficit with consequences in later

Table 1. Late complications of Fontan surgery

Organ involvement	Late complications
Bone	Osteopenia Osteoporosis
Brain	Cerebral abscess Cerebrovascular events Neurodevelopmental delay White matter changes
General	Cyanosis Delayed puberty Exercise intolerance Fertility issues Issues related to genetic syndromes Growth issues Psychological trauma
Heart	Arrhythmia – tachycardia & bradycardia Fontan pathway obstruction Outflow tract obstruction Pulmonary venous pathway obstruction Valvular heart disease Ventricular dysfunction
Immune system	Reduced immunoglobulins
Kidney	Reduced glomerular filtration rate
Liver	Cirrhosis Hepatocellular carcinoma (HCC) Liver fibrosis Portal hypertension
Lungs	Bronchial casts Hemoptysis Restrictive lung physiology
Lymphatics	Chylous effusions Plastic bronchitis Protein losing enteropathy
Pregnancy	Cardiac complications Fetal congenital heart disease Fetal loss Prematurity Hemorrhage
Vascular	Arterial and venous collaterals Dilatation of Damus Endothelial dysfunction Paradoxical embolus Venous thromboembolism

life, and there is frequently a psychological impact. A consistent long-term surveillance strategy for complications is necessary through adolescent and adult life.

Warchoł-Celińska et al. [3] carried out a multicenter study to assess the management of Fontan patients in Poland. They surveyed 8 centers caring for a total of 398 adolescent and adult patients with a Fontan circulation. Data were collected on follow-up, cardiovascular examination, and surveillance investigations. They applied a novel scoring system to create a standardized method of comparing centers. They demonstrated differences in the care and follow-up of adolescent and adult patients; with centers that provided care for pediatric patients (up until 18 years) achieving a better score.

Adolescents were more frequently followed up whilst adults were more likely to be hospitalized. The frequency of basic investigations such as electrocardiogram (ECG), echocardiogram, NT-proBNP, and chest X-ray (CXR) was satisfactory in both groups. There were, however, deficiencies in functional assessment and surveillance of long-term complications, with many patients not undergoing appropriate exercise assessment, liver monitoring, or cardiac evaluation with cardiac MRI.

The authors highlighted the need for a standardized approach to the long-term care of patients with Fontan palliation. European Society of Cardiology (ESC) guidelines for management of adult congenital heart disease (ACHD) recommend a follow-up strategy for these patients and others with complex congenital heart defects, in whom similar principles of care apply [4]. Using ESC guidance as a basis for clinical practice, we have developed a standarized approach to the transition of care from pediatric to adult care and lifelong follow-up of patients living with a Fontan circulation (Table 2).

Transitional care is vitally important to ensure long-term compliance with surveillance strategies, optimize approaches to healthy living, and assist in making realistic life choices. Whilst many pediatric cardiologists are experienced in managing adult patients, the setting of care should be within the adult hospital to facilitate routine care and the management of emergency presentations and adult events such as pregnancy and routine inpatient care.

The nature of follow-up investigations is dependent on local facilities, but routine assessments of cardiac function and end-organ involvement are necessary in all patients. We follow a strategy as described in Table 2. Using a holistic approach, we recommend all patients undergo dental checks twice per year, for mitigation of infective endocarditis risk. We discuss with women of childbearing age contraception and pregnancy routinely.

We also recommend follow-up in a patient-initiated approach and utilize a dedicated team of clinical nursing specialists, who are available for patient advice by email and telephone.

Table 2. Follow up strategy for adolescent and adult survivors of Fontan surgery

Timing	Assessment
Age 12–16	Transitional process begins with introduction to adult service: • Education around nature of heart condition • Lifestyle advice — sports, alcohol, tobacco, sexual health, drug use • Dental advice • Sports advice • Employment advice • Pregnancy and contraception Possible formal evaluation with cardiac catheterisation and optimization as necessary
16–18 years	Transition of care to adult services Full baseline assessment as below
All ages — annual	Clinic review and examination 12 lead ECG Blood investigations: • Renal, liver, thyroid function, bone profile • Full blood count, iron studies, clotting, HbAlc • Vitamin D • NT-proBNP/BNP Liver investigations: • Fibrosis assessment — enhanced liver fibrosis score/fibroscan • Liver ultrasound scan • Alpha fetoprotein Echocardiography
All ages at routine interval	Cardiac MRI Cardiopulmonary exercise testing
As required	Cardiac catheterization — pressures, pathway assessment, collaterals ± intervention as necessary Liver imaging — contrast enhanced MRI or Triple phase CT Lung function testing Prolonged ECG monitoring if symptomatic

Abbreviations: CT, computed tomography; ECG, electrocardiography; MRI, magnetic resonance imaging; NT-proBNP, N-terminal pro-B-type natriuretic peptide

In the UK, specialist centers offer outreach services to local hospitals in a network approach, to share experiences and expand care in the hospitals local to our patients. We recommend a collaborative working pattern with these centers and utilize a hub-and-spoke approach to routine and emergency care, with the hub being responsible for surgery and catheter-based interventions and supervising the care of pregnant women with a Fontan circulation. All specialized centers will have a relationship with a cardiac transplant center, with the ability to assess patients for possible cardiac transplantation.

ESC guidelines discuss minimal staffing levels required in a congenital heart disease center, including at least two consultant physicians each with a special interest in ACHD, ACHD structural intervention, and imaging. In addition, the regular presence of at least two congenital surgeons and two specialized cardiac anesthetists is recommended. The holistic care of these complex patients and families should be supported by dedicated clinical psychology services. National health systems have a responsibility to provide such safe levels of care for all complex congenital heart disease patients, including those with a Fontan circulation.

Article information

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