The benefits and long-term effects associated with single-ventricle (SV)/Fontan palliation are increasingly appreciated as the first generation of recipients with SV physiology enters their fourth decade of follow-up [1]. The suboptimal preload with systemic venous hypertension, abnormal cardiac contractility and elevated afterload have all contributed to the relentless but gradual deterioration in palliated patients with SV hearts. In the grown-up population, it is increasingly important to track late complications and deaths due to arrhythmias, thromboemboli, protein-losing enteropathy (PLE), worsening cyanosis and heart failure, and to elucidate the risk factors for them such as right ventricle morphology, higher Fontan pressure and era effects.

In this issue, Angeli et al. [2] reported their single-centre experience with 62 patients aged older than 16-years with SV physiology. It would have been beneficial if the article had provided robust data about the natural history of SV (separated from the effect of surgical treatment), the detailed management strategy of obsolete operations such as atrioventricular connection and any progress in the non-contemporary management.

Only three patients were in the natural history group. Three patients were in the palliated group (one Blalock Taussig shunt, one Waterston shunt and one pulmonary artery (PA) band). Fifty-six patients were in the palliated group (two bidirectional cavopulmonary anastomosis (BCPA), eight classical Fontan procedure and 46 total cavopulmonary anastomosis (TCPC)).

During the follow-up (mean 8 ± 9.1 years), three patients died (two with PLE post-TCPC and one with failed BCPA). Five patients (following PA banding, BCPA and atrioventricular connection (APC) each in one patient, respectively, and TCPC in two patients) with a combination of worsening heart failure, significant atrioventricular valvular regurgitation, tachyarrhythmias and PLE, underwent heart transplantation with no early mortality. Seven patients underwent TCPC conversion for worsening NYHA Class III–IV, with atrial fibrillation (AF) in five patients and conduit thrombosis in one patient (mean age at conversion: 22 years) with no early mortality. There was reported improvement following conversion, however, three patients remained in AF and two patients with PLE.

Thirty-three patients had magnetic resonance imaging for a functional and anatomical assessment, and 54 patients underwent CPET. The quality-of-life assessments (SF-36 and CHD-TAAQOL) using telephone interviews were performed in 51 patients. The actuarial freedom from death and transplantation was 95% at 20 years of age, 75% at 30 years of age and 50% at 35 years of age.

To provide the data relevant to the adults with Fontan palliation, the age selection in this paper was limited to those aged older than 16 years at the time of sampling. Considering that many such patients had surgery in an earlier era (1975–1985), it is perhaps surprising that there was no perioperative mortality. I only make the assumption that the patients’ selection was probably based on a group of patients who survived the surgical procedure with ‘very good’ Fontan haemodynamics and had freedom from death or transplantation in the first 16 years of life. I personally believe that it would be more meaningful if the starting point was the age at the time of first intervention or the number of years since the Fontan procedure. Encouragingly, there were no early surgical mortality rates for either transplantation or TCPC conversion. However, clinical experience has shown that the role of the TCPC conversion is limited because of the dwindling population of grown-up patients with haemodynamics suitable for these physiological/surgical innovations. Heart transplantation as an exit strategy remains intrinsically limited by immunologically-related issues and extrinsically limited by the lack of donor availability and surgical complexity. Other options, such as solid organ xenotransplantation, have been on the horizon for many years and sadly will remain there for many more years to come. The novel approach of mechanical-assist devices [3] to offer an appropriate energy source to overcome the paradox Fontan physiology because of systemic venous hypertension imposed with concomitant pulmonary arterial hypotension would require formal clinical trials.

Apart from the well-recognized sequelae of Fontan circulation, it is important to appreciate that normal ageing, pregnancy, acquired heart diseases and others would also have an impact on their quality of life.

Conflict of interest: none declared.

REFERENCES