The Current and Future Profile of the Adult Single Ventricle Patient Group

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Introduction The medium term outcome for patients born with single ventricle physiology has improved substantially with progress in Fontan surgery. However, deterioration in ventricular function is arguably inevitable. Medical therapies remain unproven, ventricular assist devices are expensive and transplantation is scarce and complicated in this cohort. It is difficult to address the clinical needs of this growing patient group when the size and status of the population is unknown.

Objective This study aims to determine the size and status of the United Kingdom (UK) adult single ventricle population currently and in the next decade.

Methods The number of surviving individuals (>16 years) with single ventricle physiology in a defined area of Northern England (resident population 2.9 million, 4.7% of UK) was identified primarily from the Northern Congenital Abnormality Survey (NorCAS). Conditions included double inlet ventricle, tricuspid and mitral atresia, hypoplastic left heart syndrome and others with Fontan surgery. Adult prevalence of single ventricle physiology was calculated. NYHA status was assessed from last clinical contact. The current paediatric population (5–15 years) was similarly determined. The UK population was extrapolated and population growth predicted by applying 10-year mortality, based on literature, to the defined populations. Migrants in and out of the region were excluded: extrapolation thus led to the lowest possible UK adult population estimates.

Results 46 adult single ventricle patients were identified in the NorCAS region. Principal diagnoses included tricuspid atresia (n=16) and double inlet ventricle (n=15). The majority had undergone total cavopulmonary connection (n=30). Few were >40 years (n=8). All patients over 30 years were NYHA class 2 or greater (p=0.018). Regional adult and live birth prevalence of single ventricle physiology were 2 per 100 000 adult population and 35 per 100 000 livebirths respectively. A current UK caseload of over 900 adults increasing to over 1700 patients by 2022 was estimated.

Conclusion The UK single ventricle population will consist of approximately 1700 adults at the end of the next decade. Data on functional status supports symptomatic decline over time. Detailed, multi-centre study is required to address availability, applicability and utilization of resources.