# [Outcomes in adults with congenital heart disease and heterotaxy syndrome: A single-center experience.](https://www.ncbi.nlm.nih.gov/pubmed/31617655)

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Congenit Heart Dis. 2019 Oct 16. doi: 10.1111/chd.12856. [Epub ahead of print]

PMID: 31617655

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**Take Home Points:**

* First study to assess clinical outcomes in adult patients with heterotaxy syndrome and CHD.
* The risk for early death, transplantation and co-morbidities remains high.
* Median survival of 54% at age 40.
* No congenital anatomic factors (ventricular status or HS type) were associated with reduced survival.
* Heart failure portends a worse prognosis.
* Nearly all patients had developed tachyarrythmias by age 50.
* 20% of patients experienced cerebrovascular accidents at point of last follow-up.
* In patients born after January 1985, intervention, including Fontan completion occurred at an earlier age.



***Commentary from Dr. Blanche Cupido (Cape Town, South Africa), section editor of ACHD Journal Watch:*** Heterotaxy syndrome is a condition characterized by the abnormal lateral arrangement of thoraco-abdominal organs, and frequently associated with complex congenital heart disease (CHD). Even in the contemporary era, morbidity and mortality remains high. Very little is known about this condition relating to CHD in adults with data currently being extrapolated from paediatric data.

This is a single tertiary center (Texas, US) retrospective folder review of adult patients between 1968 and 2018. They aimed to describe the sociodemographics and clinical outcomes in adult patients with heterotaxy syndrome and congenital heart disease.

Patients were excluded if they had situs inversus totalis, or without thoraco-abdominal laterality defect. The group was then subdivided based on splenic anatomy. A total of 62 patients met the inclusion criteria.

Of the 62 patients included, 29% (n=18) had heterotaxy syndrome with polysplenia (HS-PS), 46.7% (n=29) had heterotaxy syndrome with asplenia (HS-AS). The remaining patients had insufficient data to define splenic anatomy. The median age of the cohort was 22.7 years and 41.9% were female. Twelve (19.3%) were transplanted or dead – average age of this subgroup was 28.8 years. Three men had fathered children and one female with a Fontan circulation successfully carried a pregnancy to term.

Thirty-six interventions occurred in 24 patients after age 18 – 16 were cardiac catheterization lab interventions (angioplasty or stenting, device or coil placement, pericardiocentesis, Fontan fenestration, electrophysiology procedures). Eighteen surgical procedures occurred – Epicardial pacing, pulmonary artery plication/augmentation, Maze procedure, AV valve preplacement, Fontan conversion and heart transplant.

Forty-three patients had single ventricles of which 71% had Fontan palliations.

Twelve deaths occurred in total, 10 were pre-transplant. The overall transplant-free survival was 98.1% at age 20, 83.5% at age 30 and 54.2% at age 40 years. (Figure 1 below). Of all parameters, onlysystemic outflow obstruction neared statistical significance as a predictor for poor outcome. Neither ventricular status, nor HS type predicted survival disadvantage.

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Heart failure was seen in 29.8% (n=17/57). Those with heart failure by age 18 were more likely to die or receive transplantation with time to outcome of 2.6 years. Heart failure survival was 80.8%,58.7% and 31.1% at 20, 30 and 40 years respectively. (See figure 2 below)

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By age 18, 46.2% had no arrythmia, 25% had experienced tachyarrythmias, 17.3% had bradyarrhythmia and 11.5% experienced both tachy-and bradyarrythmias. Bradycardia-free survival was 33% at age 40. (Figure 3 below). Bradycardia was not associated with a transplant-free survival disadvantage compared to no arrythmia.

Tachycardia proved a survival disadvantage with a HR 6.48 (95% CI 1.41-29.75, p=0.016). The median age of tachycardia-free survival was 54%, 29.2% and 19.5% at 20,30 and 40 years respectively (Figure 3).

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Cerebrovascular accidents (CVA) occurred in 22.4% of patients (n=13), 7 occurring before the age of age 18. CVA was associated with transplant-free survival disadvantage (HR 7.97, 95% CI 1.93-32.99, p=0.004). CVA free survival was 84.3%, 54.2% and 40.6% at 20,30 and 40 years respectively (See Figure 4)

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Patients were stratified by ‘era’: Early era included those born before age January 1985, and the late era were those born after January 1985. Late era patients had earlier median age of Fontan completion (6.3 vs 15 years, p=0.002).