Objectives: To describe the management, course and outcome of late (following initial hospital discharge) rejection with acute haemodynamic compromise supported on extracorporeal life support (ECLS) in children with orthotopic heart transplantation (OHT).

Methods: Review of all children with acute haemodynamically relevant rejection requiring ECLS following OHT between 2/2002 and 10/2012.

Results: Of 197 consecutive children undergoing OHT (84 male; mean age 8.3±5.7 (range 0.1-18.8 years), 187 children survived and were discharged from hospital. Seven patients (of whom one patient had been transplanted elsewhere) presented with severe haemodynamic compromise after initial hospital discharge 15.3 months (median; range 1.6–140.4) after OHT. Mean follow-up was 5.0±3.1 (range 0.1-10.6) years. All 7 children required ECLS, two were placed on to ECLS following in-hospital cardiac arrest. Median duration of ECLS was 8 (range 5-15) days. All children survived to decannulation with one death after ECLS from sepsis 20 days after presentation. The median (range) duration of inotropic requirement post ECLS was 11 (5-27) days, the median ventilation time was 8 (7-30) days, median ICU length of stay was 14 (10-54) days and median hospitalization was 24 (19-118) days. In all patients, ventricular function normalized (FS>28%) within 10 (7-22) days. There was significant short-term morbidity; however, all survivors have a good functional status with no significant apparent neurological sequelae.

- 5 pts endomyocardial biopsy (EmBx) –> rejection ≥3 A (2 pts had EmBx with severe delay, due to haemodynamic instability )
- 2 pts positive C4d staining - suggestive of humoral rejection
- One pt positive DSAs preTx, 3 pts post Tx
- Number of HLA mismatches(mm) (3: n=1, 4:n=1, 5:n=4) comparable to overall group (42%-5 mismatch)
- 6/7 pts were not on our standard maintenance immuno-suppression regime (Tacrolimus/MMF) due to side effects, the only pt required ECLS in the recent era was highly sensitized (despite haemofiltration)

Despite described high mortality rates in the literature following haemodynamically relevant rejection, all 6 ECLS survivors are still alive after a median F/U of 5.9 yrs (0.7-9.2).

Conclusion: Acute rejection refractory to conventional treatment or presenting as haemodynamic collapse is a rare complication following heart transplantation. ECLS is a valuable rescue therapy in children with good medium-term outcome. Early referral to a tertiary center is strongly recommended, where ECLS is available and can be facilitated.