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Outcomes post Kawashima procedure and hepatic vein redirection for single ventricle palliation in left atrial isomerism with interrupted inferior vena cava.

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Objectives:
Patients with left atrial isomerism (LAI) and interrupted inferior vena cava (IVC) palliated with a superior cavopulmonary connection or Kawashima procedure (KP) have a high incidence of developing pulmonary arteriovenous malformations (PAVMs). We aimed to assess the clinical outcomes of patients with LAI following a KP. The main end points were death, requirement for hepatic vein redirection (HVR) and its the impact on oxygen saturations. We also sought to identity predictive factors for the necessity, or timing of HVR.

Methods: Retrospective data review of 24 patients with a diagnosis of LAI, interruption of the IVC and single ventricle physiology managed with a KP at a single centre between January 1990 and March 2010.

Results
21 patients had a KP, with 12 subsequently undergoing a HVR. There is diminishing freedom from death or HVR with increasing time following Kawashima particularly beyond 60 months. Significant risk factors for death or HVR included age at Kawashima where for every 1 month of age increase at time of Kawashima, the Hazard ratio of death or HVR increased by 0.02. The Cox proportional hazard regression demonstrates a reduced early risk ($p = 0.02$) for HVR or death in patients who underwent PA banding versus arterial shunt as the primary procedure. There was an increase in the hazard ratio for death or HVR in patients who had bilateral superior vena cavae.

HVR mortality was relatively high with 2 inter-stage deaths and 4 post HVR. The steepest trajectory of improvement of oxygen saturations is observed in the first 4 months after HVR after which the trajectory plateaus. Time between Kawashima and HVR does not influence the outcome for improvement in saturations. (p for interaction $=0.221$). However, those patients whose time between Kawashima procedure and HVR was more than five years, tended to have lower saturations at time of HVR (-6.8%, $p = 0.07$).

Conclusion
These findings advocate elective early HVR post KP in patients with LAI. Those with pulsatile pulmonary artery flow at first surgery could follow a more expectant course but with close monitoring of oxygen saturations to allow for timely intervention.