

# Cardiovascular Assessment of Children with Alagille Syndrome prior to Liver Transplantation: Should Hybrid MRI-Catheter have the Last Word

Bordin G, Valverde I, Cernat E, Baker A, Miller O.



Department of Congenital Heart Disease, Evelina London Children's Hospital, London  
Department of Child Health and Institute for Liver Studies, King's College Hospital, London  
Paediatric Cardiology Unit, University of Padua, Italy



## BACKGROUND AND OBJECTIVES

Alagille syndrome is associated with congenital heart disease (CHD), leading to a higher mortality rate after liver transplantation (LT). Combined MRI-Catheterisation (XMR) is a “one-stop shop” investigation to assess anatomy, cardiac output (CO) reserve by Stress MRI and for risk stratification. Work by our group (Razavi et al) has previously shown that patients who are unable to increase CO after dobutamine stress above a 40% threshold, are considered high risk or unsuitable for LT, even though the final decision is influenced by individual case discussion. We present our updated experience with a new era cohort and re-visit the conventional XMR criteria for LT.

## METHODS

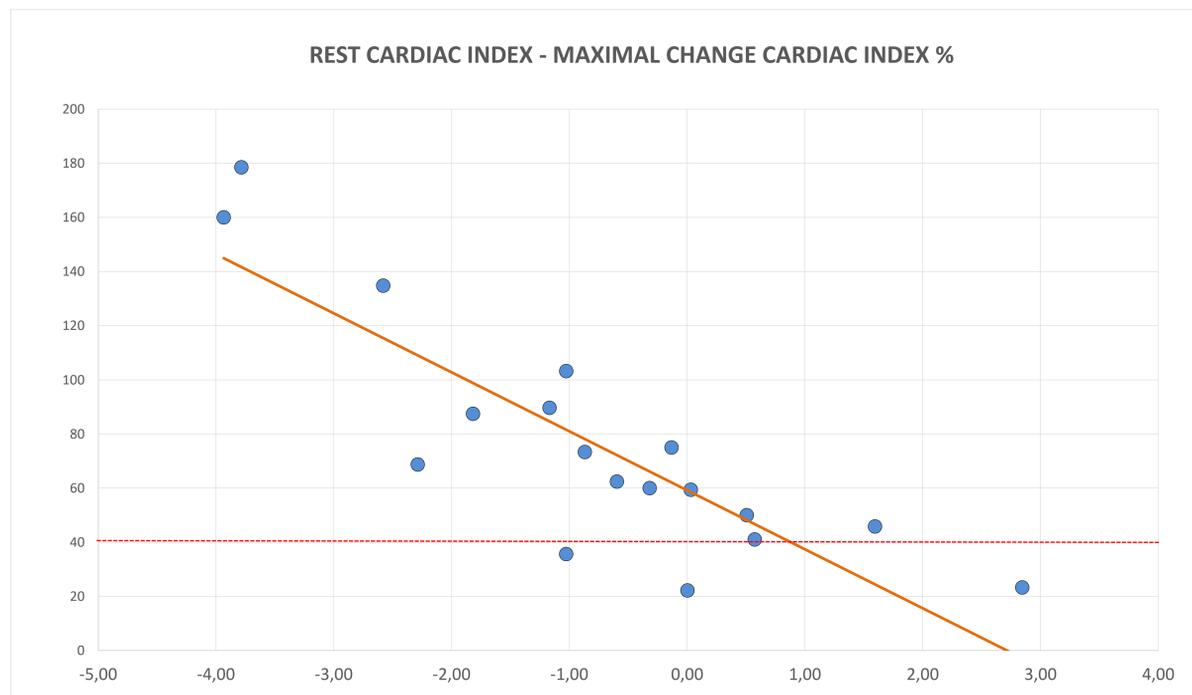
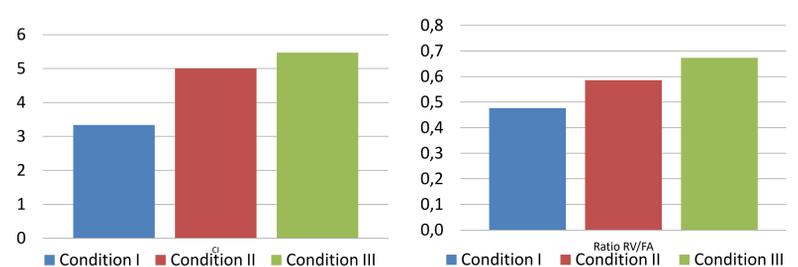
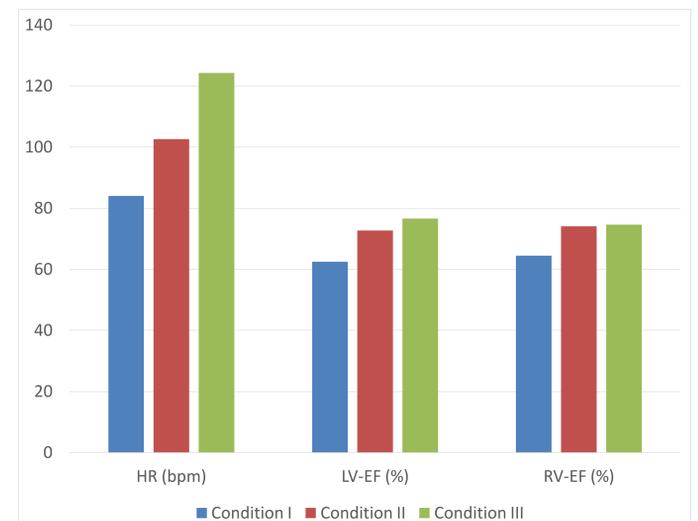
This is a retrospective single centre study. Eighteen children ( $5.6 \pm 4.3$  years) with Alagille syndrome underwent XMR at rest and during two-stages dobutamine stress (10 and 20 mcg/kg/min) between June 2007 and December 2016 in our institution.

## RESULTS

Twenty-two XMR were completed successfully in 18 children; 4 patients had 2 x XMR and one had 3 x XMR. The 18 XMR performed as pre LT assessment, showed a significant increase in heart rate and CO from baseline to maximal dobutamine stress ( $p < 0.001$ ). Three patients with significant pulmonary valvar or peripheral pulmonary artery stenosis required interventions prior to transplant consideration.

Fifteen patients were able to increase their CO more than 40% and assessed as suitable for LT: 12 patients were transplanted uneventfully, while 3 patients had stable liver status and did not eventually require LT.

Three patients did not increase their CO above the threshold of 40%. All had a CO at rest in the normal range for BMI and gender. Two were judged unsuitable for LT and subsequently died from end stage liver failure: the first patient had significant heart comorbidity, whereas the second had a normal cardiac anatomy. However, the third patient was listed as a high-risk transplant candidate, and interestingly was successfully transplanted.



## CONCLUSIONS

Dobutamine-stress XMR provides accurate information about CO response to stress, and hemodynamic and anatomic detail allowing planning of any required cardiac interventions prior to LT. It also helps to identify high risk patients, but a clear threshold cannot be confirmed from this study, and final decision still needs a case by case multidisciplinary discussion.