

Cardiovascular Assessment Of Children With Alagille Syndrome Prior To Liver Transplantation: Should Hybrid Mri-Catheter Have The Last Word

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Objectives

Alagille syndrome is associated with congenital heart disease, 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 risk stratification. Patients who are unable to increase CO after dobutamine stress above a 40% threshold, have been traditionally considered unsuitable for LT, even though the final decision is influenced by individual case discussion, as previously described by Razavi et al. 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 ± 4.3 years) with Alagille syndrome underwent XMR at rest and during two-stages dobutamine stress (10 and 20 mcg/kg/min) between August 2008 and February 2016 in our institution.

Results

Twenty-two XMR were completed successfully, 4 patients had 2 repeated XMR procedures and one had 3 XMR procedures. 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 valve or arterial stenosis required interventions prior to transplant consideration. Fifteen patients were able to increase their CO more than 40% and judged suitable for LT: 12 patients were transplanted uneventfully, while 3 patients had stable liver condition.

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 of them were judged unsuitable for liver transplant and died from end stage liver disease: the first patient had significant heart comorbidity, whereas the second had a normal cardiac anatomy. However, the third patient was listed as high risk transplant candidate, interestingly he was successfully transplanted.

Conclusions

Dobutamine-stress XMR provides accurate information of 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.