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# CONGENITAL MITRAL VALVE REGURGITATION, THE DILEMMA OF REPAIR VERSUS REPLACEMENT

## BACKGROUND

- Congenital mitral regurgitation (MR) is a rare condition and can be challenging to manage when presenting in the neonatal period.
- Medical management is preferred in order to delay surgical intervention after 1 year of life.
- When surgical intervention is indicated, mitral valve (MV) repair is preferred, however there are instances when replacement is indicated.

## CASE

- Previously healthy, full term, 2-week-old male presented with poor weight gain, murmur and cardiomegaly on chest x-ray.
- Echocardiogram showed moderate to severe MR and supra-systemic pulmonary hypertension (PHN) (figure 1, 2).
- The MV leaflets were thickened and tethered with failure of central coaptation, and the annulus was mild to moderately dilated.
- PHN was classified as WHO I and II (due to persistent PHN of newborn and MR respectively).
- Inhaled nitric oxide, Enalapril and Furosemide were initiated.
- Cardiac catheterization revealed a PVRi of 8.9 WU x m2, which was disproportionate to his degree of MR.
- A CTA was non-specific for lung parenchymal disease.
- Sildenafil and Flolan were added to reverse his PHN prior to proceeding with MV repair.
- At 4 weeks of age, he underwent mitral valvuloplasty which was complicated by severe MR and left heart failure (figure 3, 4).
- At 11 weeks of age, he underwent successful MV replacement with 17 mm St Jude mechanical valve (figure 5).
- PHN medications were weaned, and he was discharged home.

## DECISION MAKING

- This patient presented with severe left heart failure and PHN secondary to severe congenital MR.
- Medical management was unsuccessful which led to MV intervention.
- Although MV replacement can be challenging, it was ultimately necessary in this case given the severe residual regurgitation after repair.

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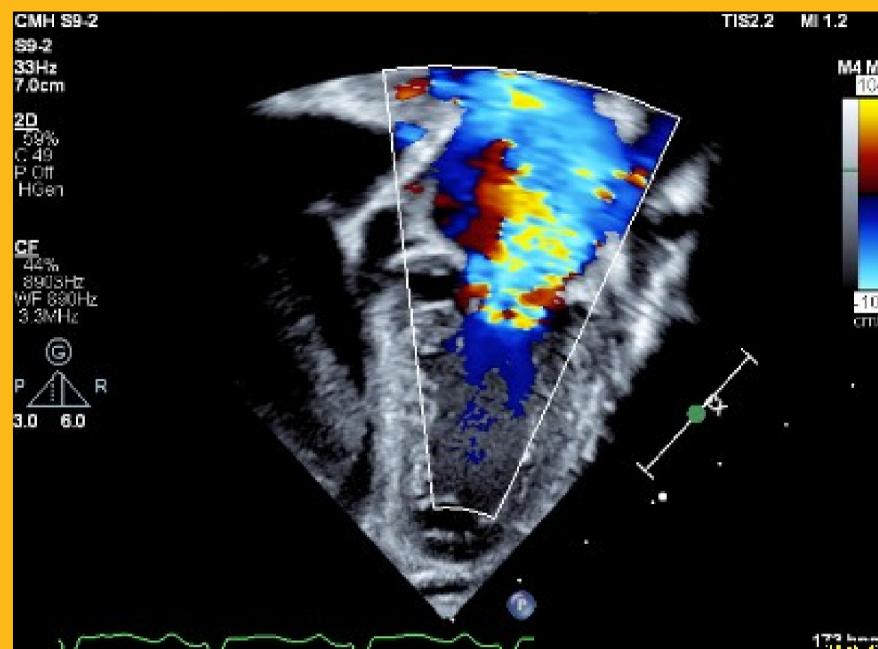


Figure 1: Apical image of severe congenital MR on presentation.

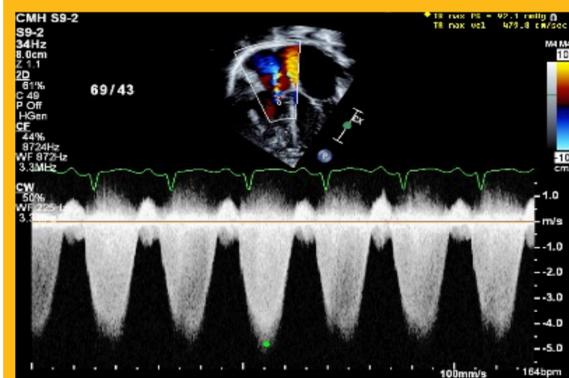


Figure 2: CW doppler of tricuspid regurgitation jet estimating supra-systemic PHN on presentation.

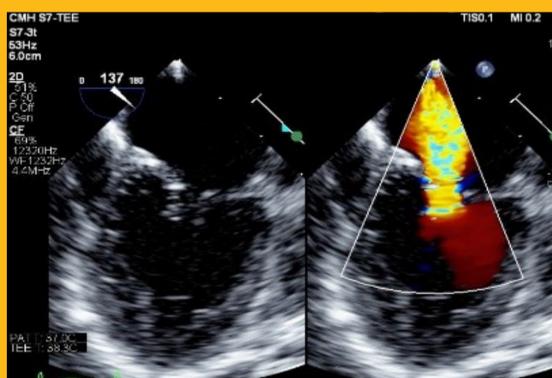


Figure 3: Post-operative TEE after MV repair showing persistent severe MR.

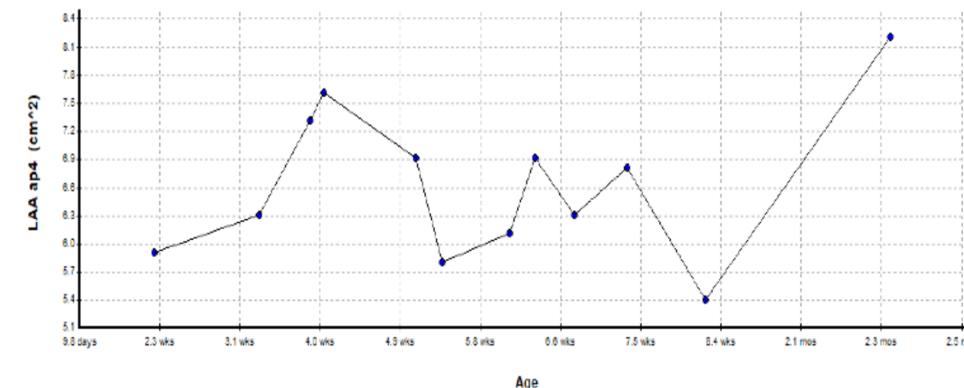


Figure 4: Graphic trends of left atrial volume prior to MV replacement.

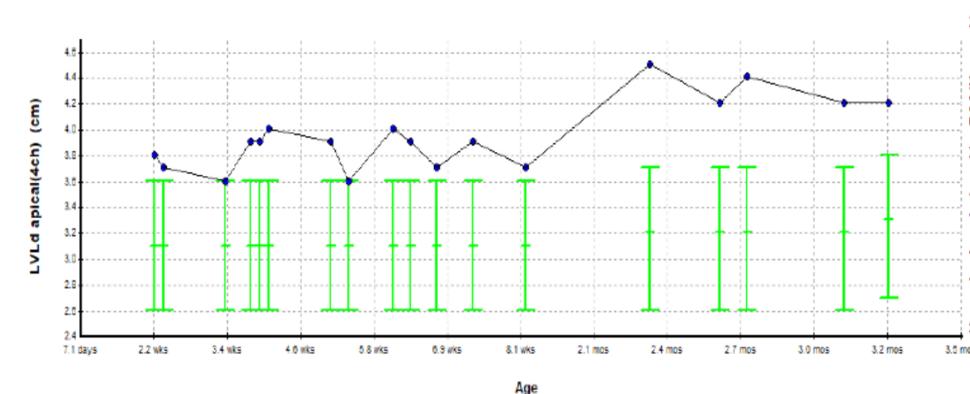


Figure 5: Graphic trends of left ventricular end diastolic volume; the last 3 data points were obtained after MV replacement.

## CONCLUSION

- This case highlights the complexity of decision making for congenital MR, and the role of MV replacement in the case of failed repair.

## DISCLOSURE INFORMATION

Neither authors have any disclosures to report.

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