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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.

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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