Guest Editorial

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Aggressive Posterior Retinopathy of Prematurity (APROP): A Preventable Challenge

Aggressive posterior retinopathy of prematurity (APROP) is a severe variant of retinopathy of prematurity (ROP)[1] which is observed in very preterm low birth weight babies exposed to poor neonatal care with unregulated oxygen and systemic risk factors. Clinically, APROP confuses beginners and experts alike, and a high index of suspicion is necessary if APROP has to be detected and treated in time to prevent poor outcomes.

APROP occurs in zone 1 or zone 2 posterior and the profile varies significantly from classical staged ROP. It has an unclear vascular-avascular junction with plus disease out of proportion to the severity seen at the junction, often accompanied by multiple vascular loops enclosing avascular areas. In severe cases involving posterior zone 1 ROP, even the macula may not be vascularized, with few vascular loops around the disc. The disease can rapidly progress to advanced retinal detachment and blindness. Therefore, as per national guidelines, ROP screening of preterm babies (<34 weeks gestation and <2000g birth weight or higher with risk factors) is very essential at 4 weeks of life (or as early as 2-3 weeks of life in very preterm babies <28 weeks gestation and <1200g birth weight) to detect ROP in time for effective subsequent management.[2]

Laser photocoagulation of the avascular retina has been a time-tested treatment modality for decades, even for zone 1 ROP. However, a trained expert is needed to laser such extensive retinal areas and also inside multiple vascular loops to minimize skip areas for effective disease regression. This extensive laser can also lead to severely constricted visual fields and high refractive errors. But the disease can be relentless and despite the best laser treatment, disease progression can still occur.[3]

The management of APROP has dramatically changed over the last few years with increased usage of intravitreal anti-VEGF drugs like Bevacizumab and Ranibizumab. These drugs have significantly improved outcomes and are now rapidly emerging as the first-line treatment of APROP. They cause fast disease regression (especially in cases with extensive neovascularization) with the added benefit that the retina continues to vascularize into the periphery, with larger visual fields and less refractive errors. They also facilitate rapid pupillary dilation allowing early fundus assessment and quick use of other treatment options like laser/surgery if required. Supported by recent results of the RAINBOW trial,[4] Ranibizumab received regulatory approval for ROP in the European Union.

The commonest clinical challenge after anti-VEGF injection is ROP recurrence at around 6-8 weeks and warrants a longer regular follow-up so that secondary laser/injection may be performed if needed. There is also limited evidence in the literature about long term safety with risks of systemic drug absorption; limited knowledge about which drug is best, how many injections to use, and what is the most effective dose to give (although half adult dose is mostly used).[5] There are new emerging challenges related to injection techniques like iatrogenic lens injury with cataract development or endophthalmitis caused due to injections in non-sterile environments. Though these drugs are expensive, support by government programs like Janani Shishu Suraksha Karyakaram (JSSK) makes them very affordable.

APROP is an important marker of the quality of care provided in the NICU. It is well known that severe ROP (and especially APROP) can be prevented if the best NICU practices are followed to prevent, minimize, and effectively manage ROP risk factors.[6] The occurrence of severe ROP and APROP requiring treatment is very low in inborn NICU babies when the high

quality of care is provided.[7] A lot of resources are needed to manage every case of APROP, and yet the outcome can be unpredictable and poor, which might lead to medicolegal issues as well. Therefore, if ROP specialists get repeated referrals for APROP from specific NICU's, it is worth discussing this with the pediatric teams, else such referrals and their resultant challenges will continue. Every effort must be made to ensure that best NICU practices are followed to prevent APROP. This will go a long way to prevent ROP related blindness.

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