Case Report

Risk Vs Benefit of Propranolol Therapy for Glaucoma Surgery in Children with Sturge Weber Syndrome

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Abstract

This article explores the use of propranolol in glaucoma surgery for children with Sturge Weber Syndrome (SWS). While propranolol shows promise in preventing ocular complications, its efficacy varies. Propranolol's mechanism involves vasoconstriction and anti-angiogenic effects but carries side effects such as bradycardia, hypotension, hypoglycaemia. We present a case of intraoperative bradycardia, emphasizing the need for heart rate and blood pressure monitoring during propranolol administration. Tailored dosing and pre-surgery discontinuation are suggested to minimize side effects. Further research is required to establish standardized propranolol guidelines for glaucoma surgery in these patients.

Keywords

Propranolol, Sturge Weber Syndrome, Glaucoma surgery, Bradycardia, Port Wine Stain

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Article History

Received: 18th November 2024
Revision: 24th November 2024
Accepted: 14th Jan 2025
Published: 27th Jan 2025

Introduction

Sturge-Weber Syndrome (SWS) is characterized by facial port-wine stain, brain angiomas and glaucoma due to abnormal overgrowth of blood vessels. The incidence of glaucoma in SWS patients is 30%–70%. Glaucoma in SWS is caused by changes in ocular hemodynamics, anterior chamber malformations, and high episcleral venous pressure because of arteriovenous shunt from episcleral haemangiomas. All these factors cause increase in intraocular pressure (IOP) leading to progressive damage. The most common uveal tract pathology in SWS is diffuse choroidal haemangiomas (DCH) that affects the posterior segment of the eyes. Macular edema and exudative retinal detachment (ERD) caused by DCH can further lead to visual impairment. Over time, untreated DCH can result in secondary neovascular glaucoma and total retinal detachment (RD).[1]

Surgical management is indicated in glaucoma refractory to medical management. However, after glaucoma surgery, early postoperative effusion of fluid into the subretinal and suprachoroidal spaces may subsequently cause ERD and choroidal detachment (CD) as reported in many cases of SWS.[1–3]

How to cite this article: Sushmita Bairagi, Renu Sinha, Kanil Ranjith Kumar Risk Vs Benefit of Propranolol Therapy for Glaucoma Surgery in Children with Sturge Weber Syndrome Ind J Ophthal Anaesth 2024;5(1):6-3

Propranolol, a beta blocker was incidentally discovered to induce accelerated involution of infantile cutaneous haemangioma (IH).[4] This discovery led to the experimental use of propranolol preoperatively for managing ERD associated with DCH in SWS. Reports have also suggested propranolol may help prevent ERD following glaucoma surgery in SWS patients.[1–3] However, beta-blockers carry significant risks, including bradycardia, bronchospasm, and hypoglycaemia.[5]

We report a case of intraoperative bradycardia in an SWS patient undergoing glaucoma surgery following propranolol administration. This highlights the potential complications of beta-blocker therapy in SWS and reinforce the need for vigilant perioperative monitoring when using propranolol for managing exudative retinal complications post-glaucoma surgery.

Description

A four-year-old male child weighing 15 kg with SWS was posted for glaucoma surgery under general anaesthesia. Pre-anaesthetic check-up was done one day prior to surgery and all routine investigations were checked. The birth history was normal and general physical examination did not reveal any systemic abnormality except heart rate (HR) of 60 beats/min with normal blood pressure and port wine staining of the face. In the treatment chart it was observed that the patient was receiving 2 mg/kg oral propranolol in two divided doses preoperatively for one week. The propranolol was started by the surgeon to reduce DCH and also decrease the likelihood of ERD post glaucoma surgery. There was no monitoring of the pulse rate and blood pressure during the administration of propranolol during the week and neither any cardiology consultation was done regarding the dose of propranolol.

Propranolol was continued till the morning of surgery. The child was taken to the operation theatre and all standard American Society of Anesthesiologists (ASA) monitors were attached. The baseline HR of the child was 62 beats/minute and the blood pressure was 100/70 mm/Hg with oxygen saturation of 100%. Inhalation induction was done with sevoflurane in 100% oxygen and after intravenous cannulation fentanyl 2 mcg/kg and atracurium 0.5mg/kg was administered. Ambu laryngeal mask airway was used for definitive airway management. Controlled ventilation was done along with boluses of atracurium and anaesthesia was maintained with oxygen air and sevoflurane with a minimum alveolar concentration (MAC) of 0.8-0.9. During surgery, patient's HR dropped to 40 beats/min and did not respond to the stoppage of the surgery. HR responded to IV atropine 0.02 mg/kg and increased to 100 beats/min which remained stable perioperatively.

Reversal of neuromuscular blockade was done after the completion of surgery and laryngeal mask airway was removed after the child was fully awake. The child was shifted to the postoperative recovery room where he was monitored for two to three hours. No drop in HR was observed in the postoperative recovery room and the child was shifted to the ward after observation. Cardiology consultation was advised in view of the intraoperative bradycardia episode and after consultation decrease in dose of propranolol to one mg/kg was advised for one week along with HR and BP monitoring.

Discussion

Sturge-Weber Syndrome (SWS) is characterized by altered vasculature, with the choroid being one of the most critical sites affected in the eye. The choroid plays a vital role in supplying nutrients to the retinal pigmented epithelium and the outer retina. Diffuse choroidal haemangiomas (DCH) are commonly associated with SWS and contribute to significant visual impairment.[1]

In patients with SWS, the primary goal of glaucoma treatment is to control IOP, thereby preventing further optic nerve damage and preserving the visual field. The most common type of glaucoma in SWS is open-angle glaucoma. To address this, surgeries such as nonpenetrating deep sclerectomy and trabeculotomy are typically recommended to create an alternative drainage pathway for aqueous humour. For cases involving malformed anterior angles, goniotomy is often employed to facilitate drainage. However, glaucoma surgeries in SWS patients carry a heightened risk of complications. These include choroidal haemorrhage, prolonged flattening of the anterior chamber, higher rates of bleb failure, and an increased likelihood of intraoperative expulsive haemorrhage during traditional trabeculectomy procedures.[1,2]

Propranolol, a beta blocker has been found to be effective in treatment of infantile haemangiomas.[4] Hence many authors have used propranolol experimentally to treat DCH and ERD in SWS.

Although the exact mechanism by which oral propranolol resolves ERD in DCH associated with SWS remains uncertain, it is hypothesized to parallel its effects on infantile haemangiomas. These effects likely include vasoconstriction, inhibition of proangiogenic factors such as vascular endothelial growth factor (VEGF), basic fibroblast growth factor (bFGF), and matrix metalloproteinases, and induction of endothelial apoptosis, ultimately resulting in vascular tumour regression.[2]

Kaushik S et al[3] observed favourable results in children with SWS with propranolol. Twelve children received oral propranolol at a dose of 2 mg/kg/day from one week before surgery till 6 weeks after surgery. No side effects of propranolol were observed and patients were benefitted by the use of propranolol in decreasing the ERD post glaucoma surgery. However no HR monitoring or baseline values were mentioned in the report.[3] Alhayaza et al reported the beneficial postoperative use of oral propranolol at a dose of 1.5–2 mg/kg/day for 4 months in a 6-year-old child with SWS.[1] In contrast, Krema et al did not find any reduction in DCH with propranolol and suggested that cavernous and capillary components of DCH in SWS may contribute to the variable response to propranolol.[2]

There is risk of bradycardia with propranolol therapy especially during general anaesthesia. Previous reports did not mention the vitals and side-effects with propranolol.

In our case, HR was on lower side preoperatively; further administration of fentanyl may have increased the risk of bradycardia during general anaesthesia and surgery. In our case, atropine was required intraoperatively for treatment of bradycardia. Resistant bradycardia with beta blocker therapy have also been reported in adults which required pharmacological intervention or even pacing.[6]

Preoperative administration of propranolol should involve consultation with cardiology and anaesthesia teams to determine the optimal dosage and timing, as its use can be beneficial postoperatively as well. Emphasis should be placed on adequate monitoring of HR and blood pressure and multidisciplinary collaboration between surgeon, cardiologist and anaesthesiologist to ensure patient safety. We suggest HR and blood pressure monitoring with lower dose of propranolol along with further research to evaluate risk and benefit of propranolol therapy to reduce DCH in glaucoma surgery in SWS patients.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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