Management of Exposure Keratopathy Associated with Crouzon Syndrome

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ABSTRACT

Introduction: Crouzon syndrome is a genetic craniofacial disorder characterised by premature fusion of cranial sutures, often leading to significant ocular complications such as exposure keratopathy.

Case: This case report discusses the management of exposure keratopathy in a 6-year-old male child with Crouzon syndrome. The patient underwent three-wall orbital decompression combined with medial tarsorrhaphy.

Observation: Post-operatively, there was a marked reduction in proptosis, better eyelid closure and significant improvement in ocular surface health.

Conclusion: This case highlights the importance of timely surgical intervention in improving ocular surface integrity and visual outcomes in patients with Crouzon syndrome complicated by exposure keratopathy. To the best of authors' knowledge, this is the first documented case of orbital decompression for Crouzon syndrome reported from Nepal.

Key words: Crouzon syndrome; exposure keratopathy; orbital decompression.

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INTRODUCTION

Crouzon syndrome (CS), the most prevalent type of craniosynostosis, is a genetic condition inherited in an autosomal dominant manner (Helmen, 2014). It is caused by mutations in the FGFR2 gene, leading to premature fusion of cranial sutures resulting in craniofacial deformities (Federica, 2025). Some features of CS are exophthalmos, hypertelorism, strabismus, exposure keratopathy, ametropia (Huang, 2024). In CS, relative proptosis from orbital hypoplasia, and underdeveloped orbital rims leads to incomplete eyelid closure. This results in exposure keratopathy, a common complication, leading to corneal drying, punctate keratopathy, and even ulceration if not timely addressed (Zeppieri, 2025). This case report outlines the management of exposure keratopathy in a young patient with Crouzon syndrome, with the use of locally available resources and a successful outcome.

CASE REPORT

A 6-year-old male child, known to have Crouzon syndrome, presented with a history of bilateral protrusion of the eyes (exophthalmos) since birth. His father had a positive family history of Crouzon syndrome. The patient had previously undergone multiple unsuccessful attempts of lateral tarsorrhaphy for exposure keratopathy in the left eye.

On examination uncorrected visual acuity was 6/12 in both eyes. The patient had noticeable bilateral proptosis, alternate exotropia and full ocular motility. In addition , the left eye showed chemosis and diffuse superficial punctate keratopathy. Both pupils were round, regular, and reactive to light. Fundus evaluation under mydriasis was normal in both eyes. Hertel's Exophthalmometry showed reading of 23 mm in Right eye and 26 mm in Left eye with bar distance of 110 mm.The diagnosis of bilateral exophthalmos with exposure keratopathy in the left eye was made, and management was initiated to address the corneal exposure. Computed Tomography scan of orbits demonstrated shallow orbits contributing to relative proptosis (exorbitism).

Given the persistence of exposure keratopathy despite previous surgical interventions, a three-wall orbital decompression (medial, lateral, and floor) of the left orbit was planned to address the exophthalmos and facilitate improved eyelid closure. A lateral canthotomy with cantholysis was performed to access the lateral orbital wall. Subperiosteal dissection was carried out to expose the lateral wall. Following periosteal elevation, the zygomatic bone and the greater wing of the sphenoid were noted to be fused which was then partially drilled using a low-cost micromotor burr and drill (Marathon Micromotor, India), creating a bony cavity while preserving a thin lateral orbital rim. Care was taken to avoid injury to the optic nerve and orbital roof. The dense lateral wall bone was progressively thinned; however, complete perforation was avoided, thereby achieving decompression while maintaining structural integrity.

The conjunctival incision was then extended inferiorly via a transconjunctival approach to access the lateral half of the orbital floor. The thickened zygomatic component of the inferior orbital wall was progressively thinned using the drill up to the lateral aspect of the inferior orbital fissure. The periorbita was incised in the lateral and inferior regions, allowing orbital fat to prolapse into the decompression cavity.

A small amount of fat was excised to enhance decompression. The periorbita was sutured to the lateral orbital wall, and lateral canthopexy was performed. Skin closure was achieved using 6-0 Vicryl sutures, while the conjunctiva was left unsutured.

A transcaruncular approach was then used to access the medial orbital wall. Portions of the ethmoid and maxillary bones were removed using a Kerrison rongeur, with care taken to preserve the nasolacrimal apparatus. The periorbita was incised to allow prolapse of orbital fat into the ethmoid sinuses, and limited fat excision was performed to further reduce proptosis. The medial conjunctival incision was left unsutured. At the conclusion of the procedure, a forced duction test confirmed no restriction or entrapment of the extraocular muscles. Haemostasis was maintained throughout, with careful avoidance of injury to the optic nerve, infraorbital nerve, and extraocular muscles.

A medial tarsorrhaphy was then performed by freshening the lid margins and placing interrupted 6-0 Vicryl sutures to partially approximate the upper and lower eyelids, thereby improving corneal protection.

Pupillary reactions were monitored hourly in the immediate post-operative period. Topical antibiotics and lubricants were prescribed, and close follow-up was advised.

At the one month, three month, and six months follow-up, the left eye exophthalmos had significantly improved, with Hertel's exophthalmometry showing a reduction of 6 mm (from 26 mm preoperatively to 20 mm post-operatively). Exposure keratopathy had fully healed, and the child reported less symptoms of ocular dryness. Functional improvements with better eyelid closure, improved ocular surface health, and enhanced visual acuity (6/9 in left eye) due to proper tear distribution was also noted. There was no change in ocular deviation after surgery.

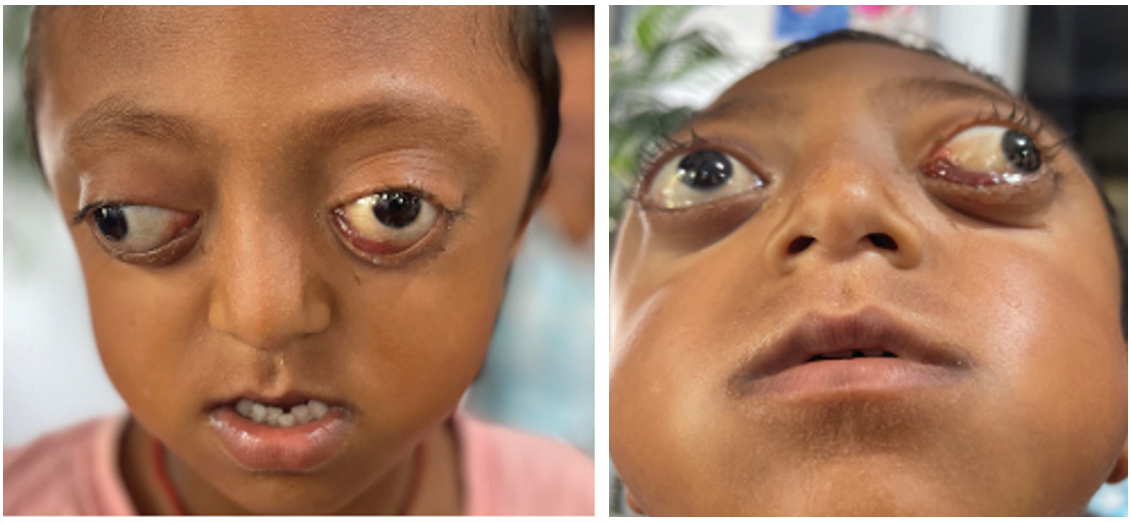

Figure 1: Preoperative picture showing bilateral proptosis, alternate exotropia, congestion, and chemosis in left eye.

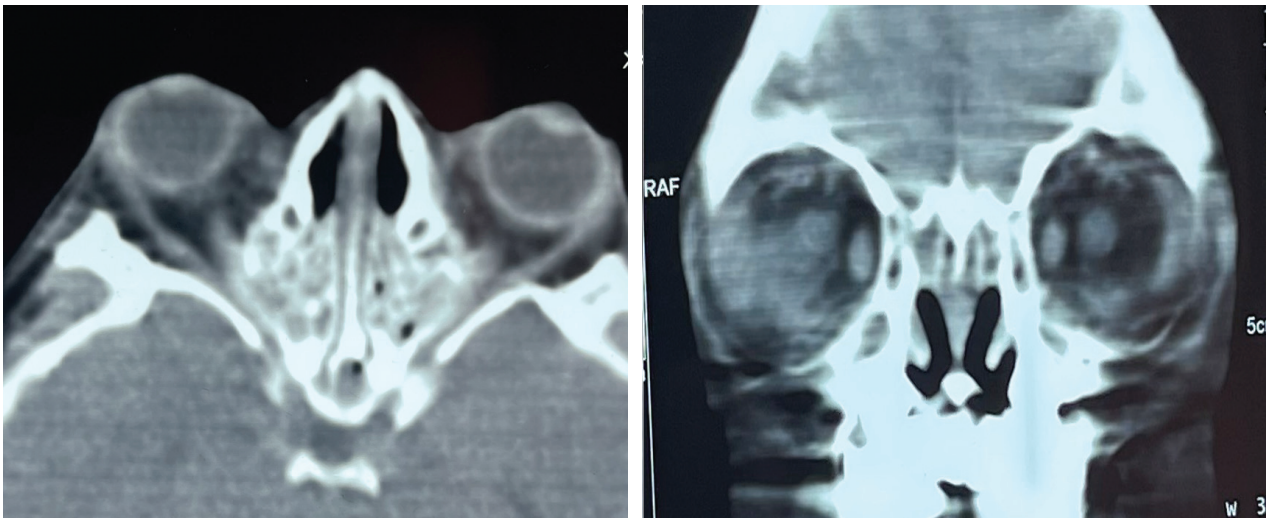

Figure 2: Preoperative Computed Tomography scan orbit (axial and coronal).

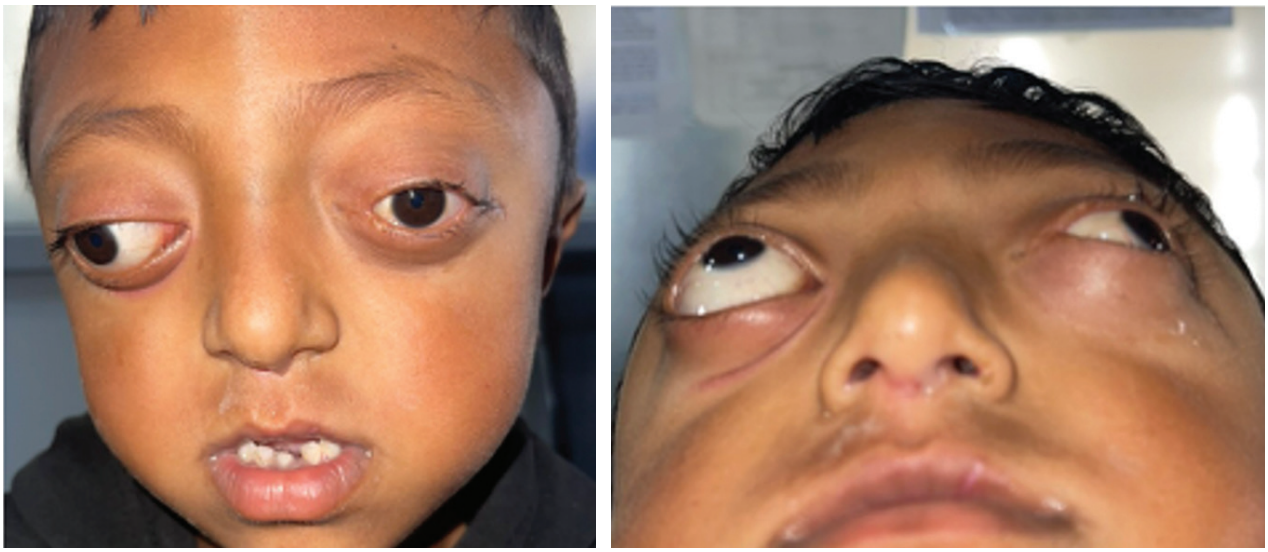

Figure 3: Three months post-operative picture of patient showing reduced proptosis in left eye, better eyelid closure and no chemosis.

DISCUSSION

Crouzon syndrome though a rare craniofacial disorder, ocular involvement is common problem. Exposure keratopathy is a frequent complication in patients with CS primarily due to exophthalmos and impaired eyelid closure. The condition can lead to corneal damage, which, if left untreated, can progress to more severe complications, including corneal ulceration and vision loss. Tarsorrhaphy is an effective measure for preserving corneal and conjunctival integrity in both acute and chronic corneal exposure, as well as in cases of conjunctival prolapse (Ganesh, 2019). But in severe case of CS with marked proptosis, lateral tarsorrhaphy alone cannot address the exposure keratopathy. In this case, multiple failed attempts of lateral tarsorrhaphy in the past highlighted the need for a more comprehensive surgical approach. The orbital crowding and exorbitism, often necessitates orbital decompression to protect the globe and improve function (Brookes, 2014).

It is often challenging to perform advanced surgeries such as orbital decompression and surgeon should have thourough experience with better understanding of orbital anatomy. Using a manual burr and drill proved effective in reducing exophthalmos and improving eyelid function in this case. The addition of medial tarsorrhaphy further supported corneal protection by enhancing eyelid closure.

Although the outcome was favourable, we recommend that, in resource-rich settings, automated ultrasonic aspirators combined with 3D facial CT guidance or image-guided navigation be utilised for more precise orbital decompression. Image-guided navigation serves as an auxiliary tool that enhances the surgeon's anatomical understanding and refines surgical precision, allowing greater accuracy in targeting specific areas, minimising complications, and optimising both cosmetic and functional results (Khan R, 2025).

There is also a reported case of endoscopic orbital decompression performed specifically for CS with severe exposure keratopathy. The procedure resulted in proptosis reduction by 3.5 mm with stable post-operative visual and motility outcomes, demonstrating its potential as a minimally invasive option in selected patients (Juniat et al., 2020).

Newer techniques like distraction osteogenesis has shown significant promise in managing severe proptosis by advancing the skeletal structure and relieving orbital pressure. This technique provides better functional improvement in eye protection by reducing exophthalmos more effectively than conventional methods. Though they come with technical complexity and long-term follow-up considerations, it represents a valuable advancement in treating cases with severe exorbitism (Bhattacharjee K et al., 2022; Hariri F, et al., 2018). A multidisciplinary approach is essential for the proper management of CS; however, financial constraints can limit access to comprehensive care.

CONCLUSION

This case demonstrates that surgical intervention can lead to favourable outcomes in managing exposure keratopathy associated with CS, even in a limited-resource setting. Despite the lack of advanced technology, proper surgical planning and techniques like three-wall orbital

decompression and medial tarsorrhaphy can significantly improve both ocular health and quality of life in these patients. With advancements in technology and surgical techniques, better outcomes can be achieved, especially in settings with more resources.

We recommend timely intervention and careful monitoring of such cases to prevent long-term complications and promote optimal visual and ocular health outcomes.

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