# Visual rehabilitation with secondary intra-ocular lens implantation in a case of Hallermann-Streiff syndrome

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## INTRODUCTION

Hallermann-Streiff syndrome (HSS) is synonymous with Francois dyscephalic syndrome, Aubry syndrome, Ullrich-Fremerey-Dohna syndrome, Oculomandibulo facial syndrome or Oculomandibulo dyscephaly with hypotrichosis.

It is a syndrome with multiple congenital abnormalities which affects face, skull, hair, eyes, teeth and overall growth and development. It affects both males and females in all ethnic groups. Over 150 cases of HSS have been reported worldwide.[1]

The potential causes of this syndrome include an asymmetric second branchial arch defect that arises during 5th or 6th gestational week, maternal viral infection, toxin exposure and paternal age. Most cases are sporadic. An autosomal dominant inheritance with variable expression or a new mutation has been mentioned, but some reports have suggested possibility of autosomal recessive inheritance.[2]

### General Features [1,2,3]

Patients generally present with small bird like facies, a beak shaped nose which is pinched and tapering at the tip. The skull is brachycephalic with frontal bossing. They have a small chin, underdeveloped jaw and a small mouth. Hair is usually sparse particularly that of scalp, brows and lashes (hypotrichosis). Skin shows atrophy. They may have natal teeth; and dental anomalies are common. Short stature is seen in about half of the individuals with HSS (proportionate dwarfism). There may be musculoskeletal and cardiac abnormalities. Most individuals have normal intelligence; however 15-30% have some degree of mental retardation. A narrow upper airway, small chin and shape of skull can pose a risk during intubation and general anesthesia. They may suffer sleep apnoea and repeated respiratory infections which can be fatal.

### Ophthalmological features [1,2]

Ocular abnormalities are a major problem with the most common ocular features being microphthalmia and cataracts which are present in 90% of HSS patients.[1] Congenital cataracts are one of the most common characteristics of HSS.[1] Cataract may be membranous or may have spontaneous absorption leading to aphakia. Other ophthalmic features include nystagmus, strabismus, blue sclera, and microcornea. Adenexal abnormalities include sparse eyelashes and eyebrows, skin atrophy, hypoplasia of lacrimal puncta, lid abnormalities (entropion, ptosis, lower lid coloboma) and down slanting palpebral fissure. Some other findings which may be present are ocular hypertension, glaucoma, pale optic disc, disc coloboma, choroidal atrophy, macular degeneration, iris atrophy, aniridia and corneal stromal opacities.

The differential diagnoses of this condition include oculodentodigital dysplasia, mandibulofacial dysostosis, cleidocranial dysostosis, progeria and other progeroid syndromes.[2]

## THE CASE

A 18-year-old Indian female patient presented with poor vision in both eyes. She was born after an uncomplicated full term pregnancy of a non consanguineous marriage. She was diagnosed to have congenital cataract at the age of one month and lensectomy was done in both eyes. She had been wearing aphakic glasses since childhood. Her general appearance and ophthalmological findings led us to diagnose her with HSS. (Fig.1) There was no notable family history. She had an average height (160 cms), and had mild mental retardation. She had microphthalmos, microcornea, nystagmus and bilateral surgical aphakia. Uncorrected visual acuity in right eye was only counting finger at 3 meters. Even with her very thick and heavy plus 16 Diopters glasses, she could hardly read the 6/60 line in the Snellen's chart. Left eye was totally blind (no perception of light, glaucomatous optic atrophy, exotropia, no fixation).

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Secondary intra-ocular lens (IOL) implantation was planned in right eye. Biometry showed short axial length (18.76mm) with normal anterior chamber depth (4.34mm). Corneal curvature measured 50 Diopters both horizontally and vertically. IOL power calculated was plus 30 Diopters. There was no posterior capsular support. She underwent glued IOL implantation under peribulbar anaesthesia. Glued IOL implantation is a technique where in posterior chamber IOL is implanted with the use of fibrin glue.[4]

Two partial thickness scleral flaps were constructed exactly $180^0$ diagonally apart followed by sclerotomy 1mm from the limbus. Thin sclera should be kept in mind while choosing the positioning of flaps. An anterior chamber maintainer was used. Anterior vitrectomy was done. Sensar 3 piece acrylic foldable IOL was injected through clear corneal incision. First, the leading haptic and subsequently the trailing haptic was externalized using an end gripping 23 G micro rhexis forceps (Micro surgical technology, USA). Both haptic tips were tucked into the intralamellar scleral tunnel made with 26G needle in the bed of scleral flaps. The sclerotomies were sutured and the scleral flaps were glued over using fibrin glue (Tisseel, Baxter, USA). Conjunctiva over sclera flaps were closed with 10-0 monofilament nylon. Corneal wound was hydrated and closed after removing the anterior chamber maintainer. At the end of surgery, well centered sclerally fixed (glued) IOL was achieved.

Post operative course was uneventful. Her vision improved to 6/36 (uncorrected visual acuity) at the end of 6 weeks. At one year follow up, vision remained the same; IOL was well centered, stable with no pseudophacodonesis; haptics were in position with no extrusion.

## DISCUSSION

A multidisciplinary approach may be required to tackle the multiple disorders of HSS patients.[3,5,6,7] Ophthalmologists have a major role to play as the patients present with multiple problems related to eyes, majority being congenital cataract or aphakia in later life (either due to spontaneous absorption of lens or surgical removal of cataractous lens).[2,3,8,9] Special attention should be paid during surgical treatment. Careful anesthetic management is needed.[1,6] IOL sizing as well as high plus power IOL could be an issue in small eyes. Shen et al have reported piggyback IOL in a patient with HSS.[2]

The short eye axis and thin sclera need cautious handling during surgery. They pose a risk of post operative hypotony, exudative retinal detachment and choroidal detachment. Cases with exudative retinal detachment after operating cataract in HSS patients have been reported.[10] Surgeon should keep in mind sclerectomy if need arises. A vitreoretinal setting should be available.

Sclerally fixed (glued) IOL is a safe and effective option for providing visual rehabilitation in patients with HSS. Good visual outcome was achieved in our patient which relieved her from aphakic glasses and significantly improved her quality of life. However, careful pre operative planning, meticulous surgery and close post operative follow up are needed.

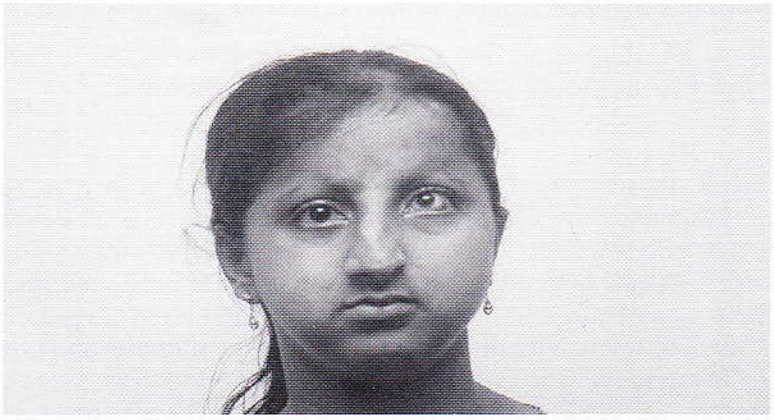

Figure 1: Bird like facies, dermatrophy of face, pinched nose, hyotrichotic patch on scalp, sparse eyebrows, mandibular hypoplasia, blue sclera, microphthalmos.

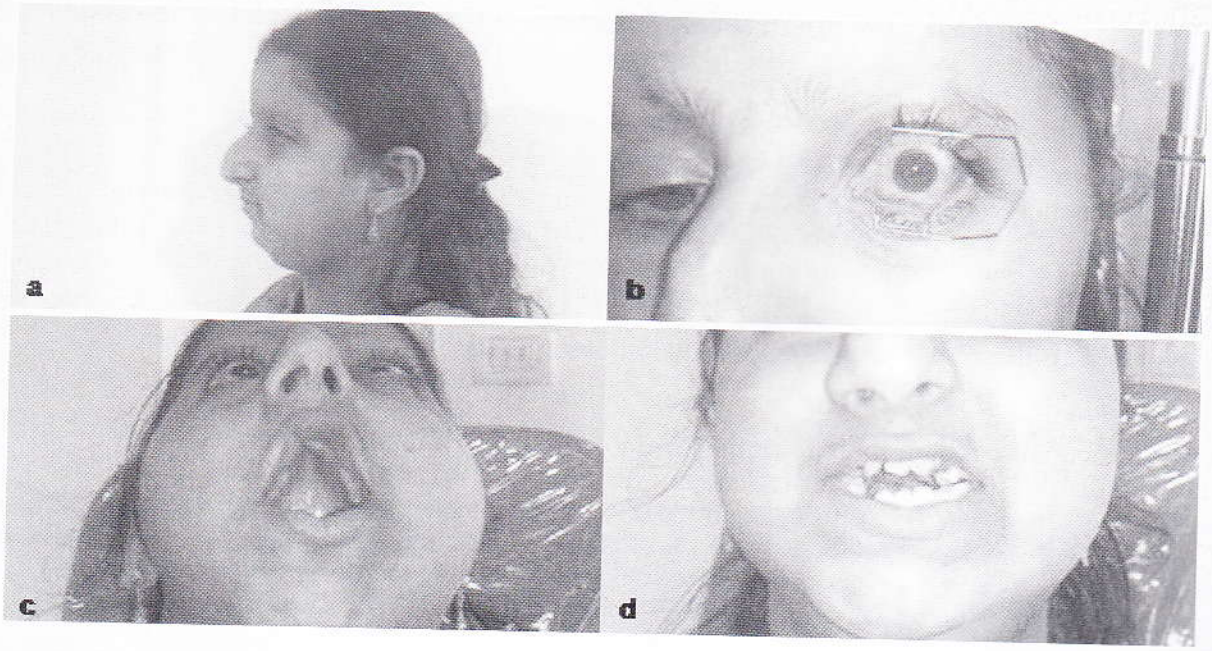


Figure 2: a) Side profile of face; b) Beaked nose, microphthalmos, blue sclera; c) Small mouth; d) Abnormal dentition, micrognathia.

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