Intact corneal sensation plays a vital role in maintaining the integrity of corneal epithelium and aids healing of epithelial defects by promoting epithelial cell proliferation.
The aim of this study is to alert ophthalmologists to congenital trigeminal anesthesia as a cause of corneal scarring and amblyopia.

Case Report
An 11 month old female infant presented to ophthalmology OPD in KKH, Najran, KSA with parents C/O Redness, lacrymation, infrequent blinking and corneal opacities.

**O/E**

The infant had delayed milestone and VA was CSM OD CUSM OS with starting nystagmus OS.

**Right Eye:**
Loss of corneal & conjunctival luster, Punctate epithelioapathy.

**Left Eye:**
Central epithelial defect, no epithelial infiltration, Conjunctival injection without much distress.
Loss of corneal sensation OU was noted as well as sensation (touch & pain) from skin of both sides of face.

Examination of both eyes was otherwise normal.

Pediatric consultation, abdominal ultrasonography as well as MRI brain were done and revealed no systemic associations.

The diagnosis of bilateral congenital trigeminal anaesthesia was reached.

Frequent application of artificial tears, tetracycline eye ointment were given with no improvement.

Punctal occlusion, lateral tarsoraphy OU and amniotic membrane grafting OS were done followed by application of soft contact lenses. Improvement was noticed.
Unfortunately, 5 days later, she rubbed against her eyes and removed the contact lens and the graft over left eye.

The amniotic membrane was rolled down into the lower fornix OS.

Corneal luster loss, punctate erosions and starting epithelial defect was noted also OD.
Re-grafting was done OU and copious artificial tears were given.

Epithelial healing was noticed and the patient had smooth course for the next 6 months.
Six months later, the patient returns C/O of redness & lacrymation OD.

**O/E**
Right epithelial defect, hypopyon, bilateral corneal opacities were noticed.

Iris, lens, fundus cannot be seen bilaterally, Loss of corneal sensation and nystagmus OU.

Fortified Ceftazidime 50mg/ml and Vancomycin 50mg/ml eye drops were given followed by amniotic membrane re-grafting and lubricants till healing of epithelium again issued.
Discussion
Congenital trigeminal anesthesia is a rare condition that was first reported by Hewsen in 1963 and defined as a congenital deficit involving all or part of the sensory component of the fifth cranial nerve (Rosenberg 1984).

The sensory deficit may occur as an isolated abnormality (as in our case), as part of a complex neurological syndrome, or it may occur in association with multiple somatic abnormalities and congenital insensitivity to pain (Rosenberg 1984).
It usually presents between the ages of 8 to 12 months with poor vision, photophobia, conjunctival injection, and corneal ulceration in the absence of pain and distress.

Punctuate keratopathy, may progress to non-healing persistent corneal epithelial defects that lead to acute corneal lysis and perforation (Ramaesh et al 2007).

Inheritance of trigeminal anaesthesia is not well defined. Most cases are non-familial and sporadic, but some cases show autosomal dominant pattern of inheritance (Purcell & Krachmer 1979).
**Stages for disease progression:**

**Stage I:**
Dry spots formation and punctate keratopathy.

**Stage II:**
Acute epithelial detachment then punched-out epithelial defect.

**Stage III:**
Stromal lysis and perforation of the cornea (Mackie 1978).

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**Differential Diagnosis:**
Among multiple causes of corneal anaesthesia neurotropic infections, such as Herpes simplex keratitis & Leprosy and the uncommon cerebellopontine angle tumor are particularly important in children (Ramaesh et al 2007).
Management of Trigeminal anaesthesia:

A) Medical:
1. Lubricants & artificial tears.
2. Bandage C L & protective glasses.
3. Anti-inflammatory agents
   - Corticosteroids.
   - Progestational steroids.
   - NSAI agents.
4. Anti-collagenolytic agents (tetracyclines).
5. Biological agents:
   - Epidermal growth factor.
   - Nerve growth factor.
   - Fibronectin.
   - Autologous serum.

B) Surgical:
1. Tarsorrhaphy.
3. Tissue glue.
4. Penetrating keratoplasty.
5. Other Techniques
   e.g. Punctal occlusion.
Our case represents a typical example of presentation and progression of an isolated case of congenital trigeminal anaesthesia that ends with corneal opacity, nystagmus and amblyopia despite relatively early diagnosis.

Conclusion
Congenital loss of corneal sensation is a rare but serious problem that should be suspected and treated as early as possible to avoid grave complications...

Thank You