Crosslinking for Keratoconus in Down syndrome

C. Koppen MD, I. Leysen MD, MJ. Tassignon MD, PhD

Antwerp University Hospital, Dep of Ophthalmology
Antwerp University, Faculty of Medicine
CXL for Keratoconus in Down syndrome

Down syndrome and the cornea

Thinning of the cornea (0.48 vs 0.55 mm) may account for a steeper keratometry (46.39 vs 43.41) and a higher frequency of (oblique) astigmatism due to lower corneal rigidity.

CXL for Keratoconus in Down syndrome

Keratoconus and Down syndrome.

This corneal thinning may also be of etiological importance to the increased incidence of keratoconus in Down syndrome: between 5 and 15%

Hypotheses:
• Genetic alterations leading to structural or biochemical changes in the cornea
• Extensive eye rubbing

CXL for Keratoconus in Down syndrome

Keratoconus and Down syndrome

Acute keratoconus or hydrops is a rare condition (2.8 %), but it is more frequent in Down syndrome patients.

In Down syndrome acute hydrops can be the presenting feature and is an important cause of blindness.

Appelmans 1961: 10 Down/52 patients.
Haugen 2001: 10/33 Down patients.

CXL for Keratoconus in Down syndrome

*Keratoconus and Down syndrome*

**Purpose:** To report on 2 cases of Down syndrome with progressive keratoconus who underwent bilateral simultaneous crosslinking therapy under general anesthesia.

To discuss keratoconus in Down syndrome as an indication for corneal crosslinking.
CXL for Keratoconus in Down syndrome

Case 1

18 year old Down syndrome young man, goes to school, is able to read, rides a horse, drives a bike.

In 2005 BCVA 0.4 OD and 0.3 OS.
In 2007 BCVA 0.3 and 0.1 OS – has lost the capacity to read with the left eye.
CXL for Keratoconus in Down syndrome

Case 1: bilateral progression
CXL for Keratoconus in Down syndrome

Case 1

Hypoosmolar riboflavin is used for both eyes because stromal thickness $< 400 \mu$, the left cornea remains at 300 $\mu$ and is not treated.

Post-CXL: bilateral bandage contact lens and eye shield, Ofloxacin 0.3% drops 4 times a day and Lorazepam 1 mg + constant supervision by the mother.
CXL for Keratoconus in Down syndrome

Case 1: fu of 2 years
BCVA OD has remained stable at 0.3
CXL for Keratoconus in Down syndrome

Case 2

17 year old boy with more limited capabilities.
Objective and subjective signs of decreasing BCVA
and steepening of the cornea.
Thickness of the stroma > 400 µ in both eyes
R/ standard CXL treatment.
Post-CXL: bilateral bandage contact lens and eye shield,
Ofloxacin 0.3% drops 4 times a day and Lorazepam 1 mg
+ constant supervision by the mother.
CXL for Keratoconus in Down syndrome

Case 2

Left eye heals uneventfully.

On day 2: right eye corneal ulcer,
culture shows growth of Streptococcus pneumoniae.

R/ hospitalisation and fortified antibiotics 24/24 h
leads to healing of the ulcer.
CXL for Keratoconus in Down syndrome

Case 2

On day 12 recurrence of the infectious ulcer in the right eye
R/ hospitalisation and fortified antibiotics 24/24 h.
It becomes clear that the mother has no authority over the son, he is rubbing his eyes, he needs a reward for every drop that needs to be instilled.
Sedation is necessary to do the follow up and treatment:
lorazepam, haloperidol, clotiapine,…
Case 2

Nothing really works: on day 15 the patient does not allow any longer examination nor treatment of the eye!

*R/ general anesthesia for examination and subconjunctival antibiotics,

followed by hospitalisation in the intensive care unit for 3 days.

*R/ tarsoraphy on day 18 before the patient regains consciousness.
Case 2

Tarsoraphy is left in place for > 6 months.
Scarring of the paracentral cornea with a final BCVA 0.6
= no loss of lines.
CXL for Keratoconus in Down syndrome

Conclusion

Progressive keratoconus in Down syndrome is a good indication for crosslinking

• to avoid visual disability and decreased quality of life
  • to avoid hydrops and scarring
  • to avoid transplant surgery
CXL for Keratoconus in Down syndrome

Conclusion

Explain to the patient and his / her family that treatment can be more challenging because of:

• lack of cooperation
• extensive eye rubbing
• self-inflicting behaviour

CXL for Keratoconus in Down syndrome

Conclusion

“Good results can be obtained in penetrating keratoplasty for keratoconus in patients with Down's syndrome who do not demonstrate a tendency toward excessive eye rubbing and for whom a single observant caretaker can be relied on to provide consistent postoperative care.”

CXL for Keratoconus in Down syndrome

Conclusion

“Good results can be obtained in crosslinking for keratoconus in patients with Down's syndrome who do not demonstrate a tendency toward excessive eye rubbing and for whom a single observant caretaker can be relied on to provide consistent postoperative care.”