Crosslinking for Keratoconus in Down syndrome

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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.

Haugen O et al. ***Biometric measurements of the eyes in teenagers and young adults with Down syndrome. Acta Ophthalmol Scand. 2001; 79: 616-625***

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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*

Skeller 1951, Cullen 1963, Shapiro 1985, Frantz 1990, Haugen 1992, Castane M 2004.

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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.

Tuft S. et al. Acute corneal hydrops in keratoconus. Ophthalmology. 1994; 101:1738-1744.
Haugen O. et al. Corneal grafting for keratoconus in mentally retarded patients. Acta Ophthalmol Scand. 2001; 79: 609-615.

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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.

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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.

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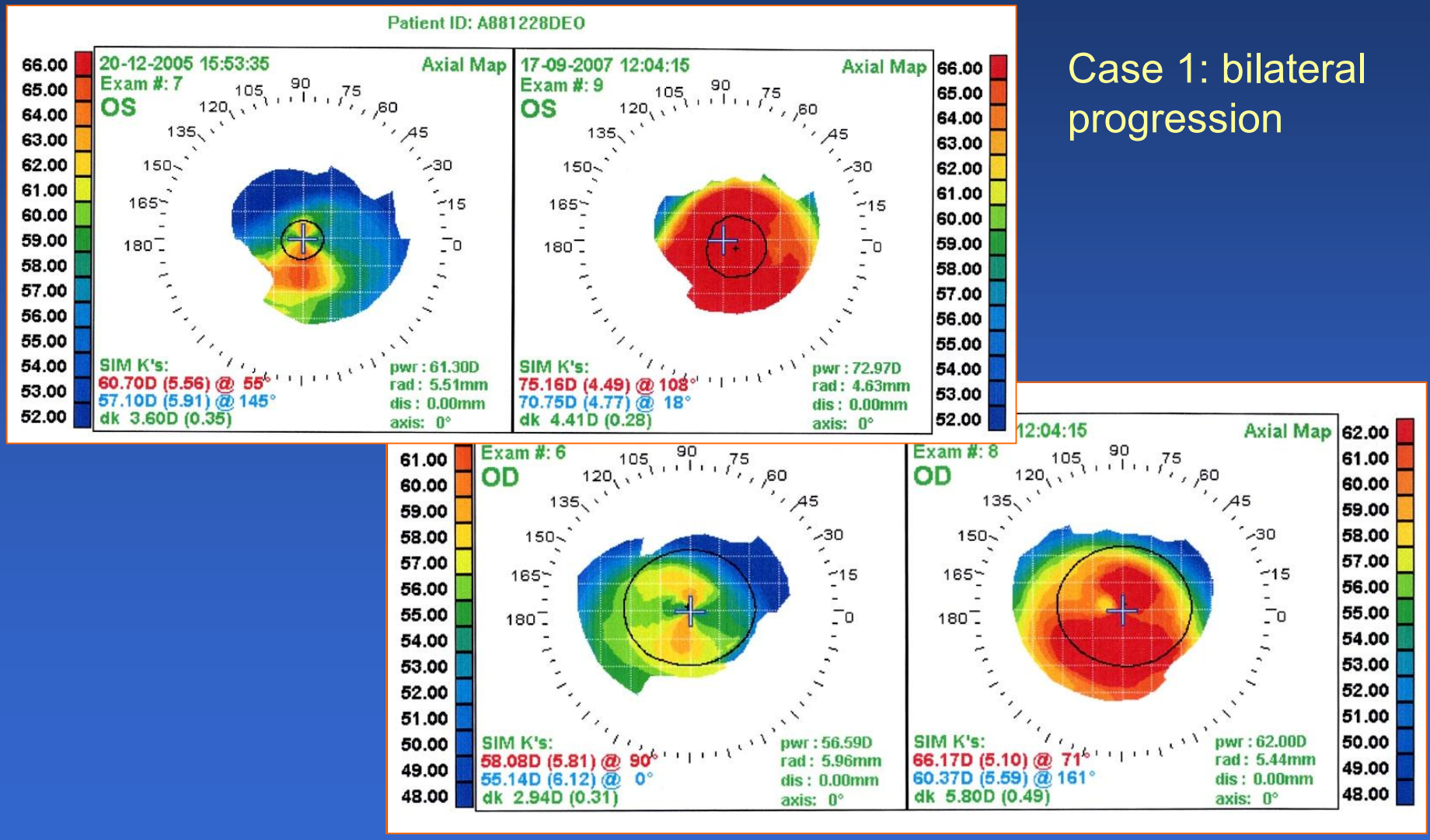

Case 1: bilateral progression

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Case 1

Hypoosmolar riboflavin is used for both eyes
because stromal thickness << 400 µ,
the left cornea remains at 300 µ and is not treated.
Post-CXL: bilateral bandage contactlens and eye shield,
Ofloxacin 0.3% drops 4 times a day and Lorazepam 1 mg
+ constant supervision by the mother.

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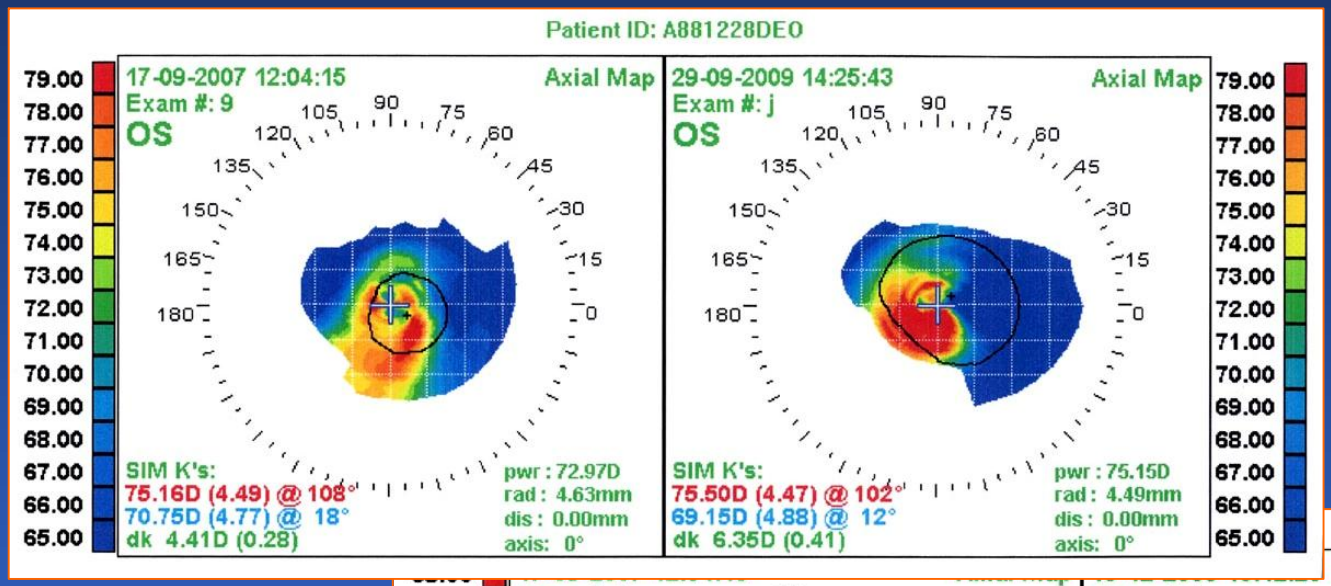

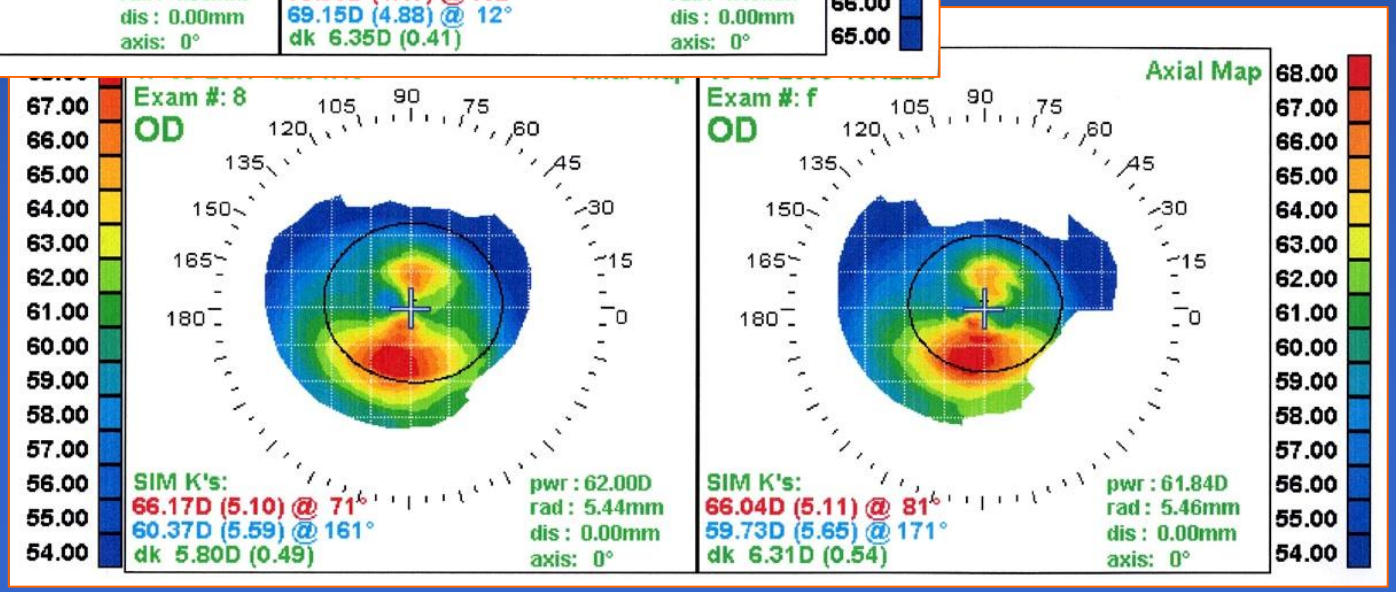

Case 1: fu of 2 years
BCVA OD has
remained stable at 0.3

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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 contactlens and eye shield, Ofloxacin 0.3% drops 4 times a day and Lorazepam 1 mg + constant supervision by the mother.

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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.

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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,…

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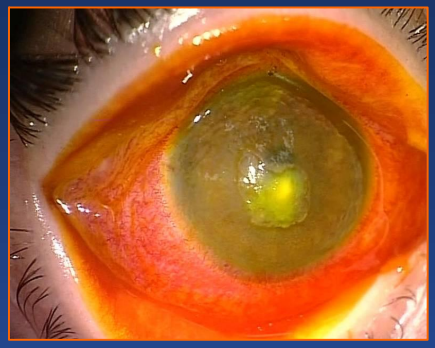
Case 2



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.

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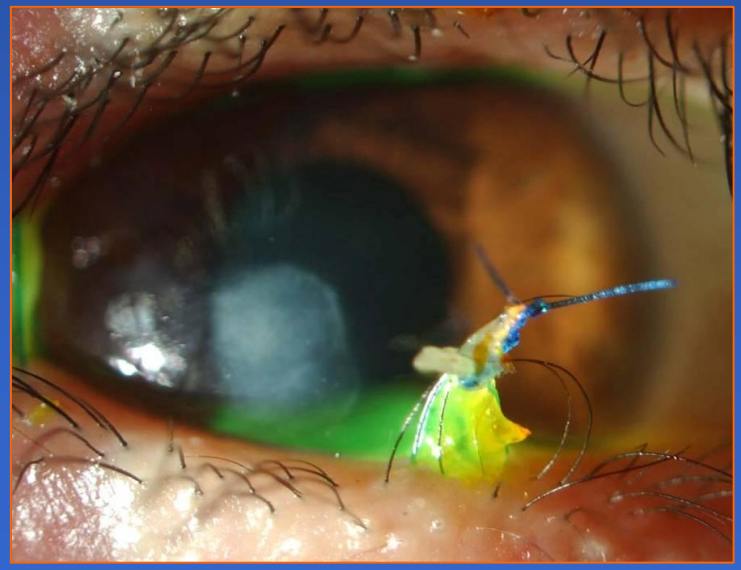
Case 2

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



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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*

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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*

Slusher 1968, Sharif 1991, Koenig 1993, McElvanney 1997.

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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."

Frantz JM et al. Penetrating keratoplasty for keratoconus in Down's syndrome. Am J Ophthalmol. 1990; 109: 143-7.

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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."