

Down Syndrome With Ectropion

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Abstract

Down syndrome is associated with characteristic eyelid abnormalities. Ectropion is one of the rare ocular manifestations of Down syndrome. We report a 4 year old boy with Down syndrome and ectropion. Ectropion in Down syndrome may resolve spontaneously or it requires surgical intervention if complication arises.

INTRODUCTION

Eye changes in Down syndrome include nystagmus, strabismus, cataract, Brushfield's spots, alternate esotropia, partial optic atrophy, retinal detachment, congenital glaucoma, ectropion and keratoconus¹. We report a 4-year-old boy with Down syndrome and ectropion.

CASE REPORT

A 4-year-old boy presented with clinical features of Down syndrome like generalised hypotonia, round facies, upward slant of eyes, ectropion (Fig1), low set ears, small mouth with protruding tongue, simian crease and clinodactyly. His cardiovascular system examination was normal. His karyotyping was 47 XY, + 21. His ophthalmic examination revealed epicanthic folds, bilateral ectropion, euryblepharon and lid retraction. There was no cataract but exposure keratitis was present. There was no retinal detachment. Visual acuity can not access as he was not cooperative, but his vision was grossly intact. His intraocular pressure was normal. The ophthalmologist advised lateral tarsorrhaphy later if symptomatic treatment with corneal lubricants, moisture shields fails. He was advised to regular follow up and surgery. But the parents refused surgery and lost follow up.

Figure 1



DISCUSSION

Though the ocular manifestations of Down syndrome are many, reports of ectropion are rare^{2,3}. It occurs due to faulty development of the lateral canthal ligament. As the supporting framework of the lid is altered, the entire lid margin falls away from the globe and the palpebral conjunctiva and fornix get exposed⁴. The complications associated with this condition are epiphora, punctal phimosis, keratinization of the lid margin and palpebral conjunctiva, chronic conjunctivitis and exposure keratopathy.

Prevention of exposure keratopathy is one of the major goals of management. Taping shut of eyelids and application of artificial tears and lubricant gels can do this. If medical management fails surgical management by cantholysis with tarsal tightening procedure at its lateral margin is done. This permits reattachment of the tarsal plate to the periosteum.

Excess skin is excised and orbicularis muscle is resutured₅.

CONCLUSION

We report this boy to create awareness among pediatricians and ophthalmologist about the eye findings in Down syndrome. Congenital ectropion in Down syndrome may resolve spontaneously or may need surgical management to avoid exposure keratopathy.

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References

1. Liza-Sharmini AT, Azlan ZN, Zilfalil BA. Ocular findings in Malaysian children with Down syndrome. *Singapore Med J* 2006; 47: 14-19
2. Sellar PW, Bryars JH, Archer DB. Late presentation of congenital ectropion of the eyelids in a child with Down syndrome: a case report and review of the literature. *J Pediatr Ophthalmol Strabismus*. 1992; 29: 64-7
3. Miller R, Martin F, Allen H. A case of congenital ectropion in Down's syndrome. *Aust N Z J Ophthalmol*. 1988; 16: 119-25
4. Alvarez EV, Wakakura M, Alvarez EI. Surgical management of persistent congenital eversion of the upper eyelids. *Ann Ophthalmol*. 1988; 20: 353-4
5. Morris RJ, Collin JR. Functional lid surgery in Down's syndrome. *Br J Ophthalmol*. 1989; 73: 494-7

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