

Unilateral congenital buphthalmos

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DESCRIPTION

A term female infant was born by forceps delivery after failed ventouse. There was a history of maternal fever and antenatal antibiotics. She was in good condition after birth but was noticed to have an 'enlarged and cloudy right eye'. Elevated C reactive protein led to sepsis investigations and giving the infant intravenous antibiotics. Blood culture and cerebrospinal fluid microscopy and culture were all negative. Brain MRI revealed no evidence of haemorrhage, ischaemia or space-occupying lesion.

Expert review by the ophthalmologist established the diagnosis of diffuse right corneal oedema and buphthalmos (figure 1). Clinical examination of the left eye appeared normal (figure 2). Digital and rebound tonometry with iCare showed intraocular pressure over 25 mm Hg in the right eye and less than 15 mm Hg in the left eye. The differential diagnosis included congenital glaucoma or anterior segment dysgenesis, but the possibility of a rare congenital corneal dystrophy or other underlying ocular pathology was also considered. Pilocarpine eye drops were introduced with good response in terms of corneal oedema and intraocular pressure. Despite the unilateral nature of the disease, the metabolic screen sent for urine amino and organic acids, galactosaemia and lysosomal neurodegenerative and storage diseases, revealed no obvious abnormalities. The infant's surgical treatment started at 2 months of age.

Buphthalmos (*greek for "Ox Eye"*) can be the result of congenital glaucoma, and without timely

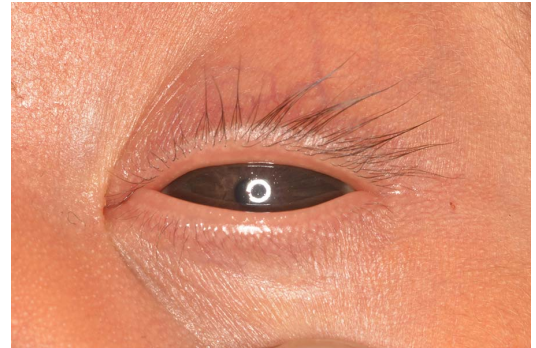


Figure 2 Normal left eye.

medical and surgical treatment may lead to seriously affected vision.¹ Unilateral congenital corneal oedema may be secondary to congenital glaucoma, anterior segment dysgenesis, neurodegenerative/storage diseases, neurofibromatosis, or birth injury by forceps.¹⁻³

Learning points

- ▶ Buphthalmos due to congenital glaucoma may be unilateral.
- ▶ The condition requires prompt review and management by senior paediatricians and ophthalmologists, and referral for surgical treatment is required in the majority of cases.
- ▶ Aetiology includes congenital abnormalities, neurometabolic diseases and trauma.

Competing interests None declared.

Patient consent Obtained.

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Figure 1 Diffuse right corneal oedema and buphthalmos.



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