Spontaneous luxation of the eyeballs

Report of a patient with brachycephaly and anomalies of the extraocular muscles

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A study of the medical literature of the past 70 years revealed no fewer than 46 cases of spontaneous or voluntary luxation of the globes. While we cannot uphold our earlier impression that this is an excessively rare condition, it would seem sufficiently uncommon for an ophthalmologist to be unlikely to encounter more than one example in his professional lifetime.

Fowler (1941) credited Willemer (1892) with the first report of the condition. In a comprehensive review of the reported examples, Fowler distinguished between luxation, dislocation, and avulsion of the globes. Luxation means that the eye protrudes between the eyelids with spastic closure of these behind it. Dislocation signifies traumatic displacement of the eye into the nasal sinuses or nasal cavity. Avulsion implies that the extraocular muscles and optic nerve have been partially or totally severed from the body. Luxation (forward displacement of the eyeball) may be spontaneous or voluntary. Spontaneous luxation has occurred as a consequence of exophthalmic ophthalmoplegia with extreme exophthalmos (Sattler, 1909; Miller, Schlossman, and Boyd, 1947), shallow orbits in craniofacial dysostosis (Donaldson, 1903; Franceschetti, 1939), oxycephaly (Fletcher, 1911), brachycephaly (Fowler, 1941), retraction of the eyelids to remove a foreign body (Galloway, 1910), excess orbital fat (Offenbach, 1954), tumour (Nicati, 1951), and syphilitic gumma (Tucker, 1907). Voluntary luxation of the globes occurs mainly in Negroes (Lyle and McGavic, 1936), and has been attributed to a combination of shallow orbits, lax ligaments (Almeida, 1932), and reduplication of the superior oblique muscles (Gillespie, 1964). One patient was able to cause luxation by blowing the nose violently (Ball, 1926). Sometimes voluntary luxation has been assiduously developed and practised by the individual for use as a trick (Ferrer, 1928: quoted by Waardenburg, Franceschetti, and Klein, 1961), or as a means of earning a living (Maskelyne, 1895: quoted by Foster, 1952).

The exact aetiology of these cases was not always established with certainty and skull $\times$ rays were often omitted. The availability of skull and orbital tomograms has allowed us to make detailed measurements of their dimensions and to diagnose brachycephaly in this patient with some confidence, and as the patient described by Fowler (1941) seems to be the only other recorded in the literature with this complication, we thought that ours would be of interest.

Case report

On October 16, 1965, a colleague asked one of us (E.M.) for an urgent consultation on a patient he had been following for about 30 years. She was 72 years old and a diabetic. About 7 years pre-

viously she had had her first episode of luxation of the right globe. In the last 18 months she had had more frequent episodes of luxation of the right eye and occasionally of the left. In the last few weeks, these attacks had become much more frequent, harder to reduce, and painful. The frequency of the luxations of the right globe carried with them the risk of interruption of the blood supply by strangulation and blindness from optic atrophy. The situation was made worse by the fact that the left eye was amblyopic and the patient used the right eye for fixation. Another ophthalmologist had recently attempted to prevent luxation by performing a right lateral tarsorrhaphy. The result of this had been to aggravate the existing ptosis of that eye. The right cornea was totally occluded and the patient became effectively blind. She was unable to go about her business and the lateral tarsorrhaphy had to be opened immediately.

She was immediately admitted to the Ophthalmic Foundation in Paris. By this time she was luxating her globes spontaneously without any provoking factor, whereas previously this only occurred when she tried to open her eyes or wash her face or when she was examined by an ophthalmologist.

Examination

She was a reasonably well-nourished woman. There was a band-shaped opacity of the left cornea. The left eye was amblyopic and showed a secondary divergent strabismus (Fig. 1). Ptosis was present. She was fixing with the right eye and this had a paracentral corneal nebula. The visual acuity of the right eye was initially 6/9 with -7 D sph., -2.5 D cyl., axis 105° . (Between 1965 and 1968 the visual acuity dropped to 6/36 as the result of a progressive nuclear and posterior cortical cataract). She was able to read No. 3 on the Parinaud reading chart without correction. The best visual acuity in the left eye was 6/60 with -2.25 D sph. An unusual feature was her oval-shaped corneae, which had vertical diameters of 12 mm. and horizontal diameters of 10.5 mm.

Figs 1 and 2 show frontal and lateral views of the patient's face. The prominence of the globes is striking and the ptosed upper lid is surmounted by the hatchet-blow appearance (*coup de hache*) which is seen after an enucleation. In both situations the levator palpebrae superioris is at a mechanical disadvantage and cannot act properly.

FIG. 1 Frontal view before operation

FIG. 2 Lateral view before operation

The lower lid of the right eye had fallen down and the equator of the globe was stretching its free border. The eyelids of both eyes were atrophic and the corneae could be seen through them like bluish rings. The sclerae were thinned and the ciliary bodies easily visible.

In addition to the bilateral ptosis, ocular motility was severely limited. Adduction and convergence were absent. Elevation and depression were practically non-existent, and abduction was very slight. The binocular field of vision (as tested with both eyes open and permitting unrestricted movement) was about 10° upwards, 10° downwards and about 20° to the right. Her history that she had always required to turn her head in order to see to the side throughout her life supported these observations.

Fundus examination showed no abnormality of vitreous or retina on either side.

Past History

She had had very prominent eyes since childhood. A photograph taken in 1917 at the age of 24 years showed this, as did a 1942 identity card photograph.

Treatment

A lateral tarsorrhaphy was performed on admission in order to protect the cornea of the right eye while various observations were made. A left lateral tarsorrhaphy had been previously performed.

TONOMETRY

Maklakov Applanation Tonometry (5 g. weight) Right eye 18 mm. Hg, left eye 18 mm. Hg.

Schiötz Tonometry on December 8, 1967 (2 years after admission) Right eye 34.5 mm. Hg (5.5 g. weight) and 29.4 mm. Hg (10 g. weight). Left eye 20.6 mm. Hg (5.5 g. weight) and 19.6 mm. Hg (10 g. weight). This discrepancy in the readings in the right eye would be in keeping with high myopia on that side and suggested reduced scleral rigidity.

Goldmann Applanation Tonometry Right eye 25 mm. Hg, left eye 17 mm. Hg. The applanation reading for the right eye was 32 mm. Hg before the instillation of pilocarpine eye drops.

GONIOSCOPY

The right eye revealed a wide open angle through 360°. The left eye had a drainage angle which was medium open in its entirety, except for several peripheral anterior synechiae between 1 and 2 o'clock.

EXOPHTHALMOMETRY (Hertel exophthalmometer)

Right eye 26 mm.—107 mm.—25 mm.

ECHOGRAPHY (ultrasonic)

The anteroposterior lengths were right eye 22.5 mm. and left eye 22 mm. measured from polaroid photographs of the oscilloscope screen.

CALIPER MEASUREMENT OF EQUATORIAL DIAMETER

This measurement, which was carried out under general anaesthesia, was 32 mm. (both eyes).

Description of surgical techniques

On December 7, 1965, an extension was made to the existing right lateral tarsorrhaphy and the lateral thirds of the upper and lower lids were united in order to protect and support that eye. The left upper eyelid covered the eye as a result of the ptosis and also of the lateral tarsorrhaphy which had been performed before her admission. An attempt to resect the inferior rectus muscle was not successful because this muscle could not be found. Instead, there was a thickened area of Tenon's capsule and this was shortened 10 mm. and sutured to the sclera. The result was to bring the pupil into the palpebral aperture and this gave the patient some peripheral field of vision.

On December 12, 1965, an attempt was made to resect the right inferior rectus muscle. We found the same situation as existed on the other side, that is, no inferior rectus. We therefore resected Tenon's capsule and attached a tongue of this to the sclera about 7 mm. posterior to the limbus. This produced an appreciable lowering effect on the cornea.

On January 7, 1966, we carried out a plastic procedure to correct the surgically-induced ptosis of the right upper eyelid (Fig. 3, opposite). This appeared difficult since it was felt that operations which depended on shortening the levator palpebrae superioris (Blasowicz, Everbusch, Iliff) would only have served to convert surgical ptosis to lagophthalmos, with recurrence of the dislocation of the globe because the tissues were generally for the same

FIG. 3 *Post-operative ptosis following extended right-sided tarsorrhaphy*

reason atrophic and the orbicularis was weak. The method of fixation of the tarsus of the upper lid to the frontalis muscle by a silk suture described by Friedenwald and Guyton (1948) was considered and rejected. In the end we decided to modify the V-Y operation for ectropion (Wharton Jones) described by Stallard (1958) and to convert it into a Y-V procedure for the upper lid. An upside down Y was cut in the skin of the upper lid and the vertical portion was continued upwards, cutting the eyebrow, to a point 5 mm. above it. The skin from the angular part of the Y was dissected clear and a V-shaped fold was raised and sutured to the upper extremity of the vertical incision. The elasticity of the skin allowed a satisfactory suspension of the upper lid and cleared the pupillary area. At the same time, it allowed the palpebral part of the orbicularis muscle to work and prevented any further luxation of the globe. It must be admitted that our earlier attempt was too conservative and the operation had to be repeated in order to give a satisfactory result.

On September 16, 1966, a Y-V procedure was carried out with the same intention on the left upper lid. This first operation was inadequate and had to be repeated a month later. The final result was functionally satisfactory on both sides and aesthetically less unsightly than had been foreseen (Figs 4 and 5).

FIG. 4 *Lateral view, showing right eye following extended lateral tarsorrhaphy and Y-V plastic procedure*

FIG. 5 *Frontal view, showing appearance after extended lateral tarsorrhaphy and bilateral Y-V eyelid elevation procedures*

Meanwhile the chronic glaucoma was kept in check by pilocarpine with the disadvantage of miosis on the nuclear and posterior cortical cataract affecting the right eye. The risk of vitreous loss from a globe with a low scleral rigidity inadequately supported by the orbit which was tantamount to a bony plateau, made us postpone cataract extraction as long as possible. Our first surgical approach to this problem was the performance in February, 1968, of a three-snip optical iridectomy which allowed the unimpeded use of miotics in the right eye.

By June, 1968, the visual acuity had fallen to below 6/60 and operation became imperative. On June 18, 1968, under basal analgesia, using diazepam (Valium) and a neuroleptic agent, a cryoextraction of the lens with peripheral iridectomy was carried out on the left eye. Acetazolamide was used together with the Flieringa ring and there was no vitreous loss.

On July 19, 1968, cryoextraction of the right lens was carried out by identical methods and this passed without mishap.

On July 26, 1968, one month after the lens extraction, an iris prolapse developed on the temporal side of the left eye. This was resected, and the wound was resutured and recovered with conjunctiva. There followed an uneventful recovery.

On November 6, 1968, the visual acuity of the right eye was 6/12 part with +8 D sph., + 1 D cyl., axis 170°. With a +4 D sph. reading addition she was able to read No. 2 on the Parinaud reading chart.

The intraocular pressure measured by applanation tonometry was 21 mm. Hg in the right eye and 18 mm. Hg in the left.

The patient does not use miotics and she can now lead a normal life.

Radiological aspects

Fig. 6 is a straight lateral x ray of the skull showing both orbits. These appear to be reduced in their antero-posterior diameter because of shortening of the anterior fossa.

FIG. 6 *Lateral view of skull (straight x ray) showing shallowness of orbits, which are diminished in their antero-posterior diameter*

Fig. 7 is a tomogram of a Hirtz projection at the 12 mm. setting. It shows the floor of the orbit viewed from above and an abnormally obtuse angle between the medial and lateral walls of the orbit. Table I compares these figures with the normal as given by Last (1968).

FIG. 7 Tomogram of Hirtz projection (12 mm. setting), showing floor of the orbit and abnormally obtuse angle between medial and lateral walls

Table I Comparison of patient's orbital axes with normal values (Last, 1968)

Orbital axis	Right	Left
Patient	58°	57°
Anatomical normal	45°	45°

Table II Comparison of vertical height and width of maxillary sinus with normal values (Lusted and Keats, 1959)

Measurement (mm.)	Patient				Normal
	Right	Left	Corrected by factor 1/1.3		
			Right	Left	
Vertical height	60	60	46	46	20-21
Width	49	46	38	35	19-21

Fig. 8 (overleaf), a tomogram of postero-anterior projection, shows the maxillary antrum and the ethmoidal sinuses. It shows that the orbit is being encroached upon from below and medially on both sides by very considerably enlarged maxillary antra, with the result that the floors of the orbits are very oblique inferiorly and nasally. Table II shows the numerical comparison of the dimensions of this patient with those published values of Lusted and Keats (1959). The correction factor is to compensate for the radiological magnification obtained. It is applicable here but not in Table I (Fig. 7), in which an angle and not an isolated measurement of length is considered.

Lusted and Keats (1959) defined the cephalic index as:

$$\frac{\text{The greatest transverse diameter in the postero-anterior position}}{\text{The distance between the glabella and the opisthocranion}} \times 100$$

The cephalic index in this patient was 88.4. Any value over 80 indicates brachycephaly and the 10 per cent. enlargement in this patient is thought to indicate shortening of the skull of this type.

FIG. 8 Tomogram of postero-anterior projection (19 mm. setting) showing maxillary antrum, which is enlarged in its vertical and horizontal diameters

Drey's index (1957), quoted by Lichtenberg (1960), is defined as follows:

$$\frac{\text{The vertical distance between the summit of the nasal bone and the hard palate}}{\text{The horizontal distance between the summit of the nasal bone and the anterior sellar tuberculum}}$$

The average value at birth is 0.48 and this increases to 0.67 by the age of 8 years. The value of 1.2 in this patient indicated relative shortening of the anterior half of the skull.

Discussion

Brachycephaly is a type of craniostenosis, and while several patients with spontaneous luxation have been reported, brachycephaly has been mentioned only once (Fowler, 1941). In our patient skull deformity was not obvious and the diagnosis was made from radiological measurements. Fowler noted that the skull in his patient was clinically deformed and that this was "definite" radiologically as shown by flattening in the occipital region, antero-posterior foreshortening, and shallowness of the orbital cavities.

In our patient the two commonest types of craniostenosis (oxycephaly) (Duke Elder, 1964) and craniofacial dysostosis (Crouzon's deformity) were absent. Thus there was no tower-shaped skull with flattening of the forehead nor was there any appearance of a "parrot's beak nose" with frontal bossing, as is typical of the two conditions respectively. Only a limited skull deformity was present and this was shown by the shortened aspect of the anterior cranial fossa in the lateral x ray (Fig. 6) and by the large Drey's index. This latter indicates foreshortening of the skull. It would appear that the skull sutures fused early in a limited skull area in this and in Fowler's patient.

Spontaneous luxation of the globe was caused by the fact that it was relatively too large for the orbit. Although myopia was present, echography showed the antero-posterior length of each eye to be normal. The bilateral absence of the inferior recti together with weakness of all eye movements suggests that muscle anomalies were contributory.

The transparency of the eyelids is intriguing. The cause may well have been pressure atrophy of the eyelids by the globes. This pressure on the globes by the eyelids made cataract extraction hazardous in view of the reduced scleral rigidity on the right side and the lack of support for the globes by the orbital walls. A lateral canthotomy was eschewed because we feared further luxation and loss of what had been gained at the previous six operative sessions. The use of acetazolamide and a Flieringa ring became obligatory. The availability of basal analgesia using a neuroleptic agent and diazepam was an advantage.

Summary

(1) The clinical findings in a patient with bilateral spontaneous luxation of the globe are described. The inferior recti were absent and there were probably other anomalies of the extraocular muscles.

(2) Brachycephaly was diagnosed from measurements made on orbital tomograms, including Hirtz projections.

(3) The use of Y-V plastic procedures corrected the spontaneous luxations. The management of coexistent chronic simple glaucoma and cataract in a diabetic patient with low scleral rigidity and non-existent orbital support is described.

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