Congenital total eversion of the upper eyelids

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SUMMARY Eight cases of bilateral and 6 cases of unilateral congenital total eversion of the upper eyelids are reported here. It is suggested that an appreciable overlapping of the lower eyelid margin by the upper eyelid is a predisposing factor in the mechanism of eversion in this condition. Treatment by injection of hyaluronidase into the chemotic conjunctiva followed by placement of a lid suture to revert the eyelid resolves the condition in 1 or 2 days.

Rarely babies may be born with total eversion of the upper eyelids and chemosis of the upper palpebral and conjunctival fornix. The conjunctival fornix may prolapse in severe cases. This rare condition was first described by Adams in 1896. Most of the cases reported have been in black babies. Duke-Elder stated that the condition is always bilateral, but Abiose has reported a unilateral case. Six unilateral cases are reported in this series.

The condition may rarely be associated with mongolism or colloidion skin disease. Otherwise the babies have been normal and healthy. Delivery had been normal in most reported cases, but a few authors reported prolonged and difficult labour. The aetiology of the condition is obscure and recovery rather protracted. Duke-Elder suggested that the condition is brought about mechanically as the head traverses the birth passages. He further stated that, if the eyelids are kept in their normal position for some days by adhesive strips or by sutures, and infection prevented by topical antibiotics, the lids become normal within 2 or 3 weeks.

A clinical study of the condition was undertaken: (1) to find out if there is any anomaly in the eyelid which may predispose it to eversion, since the condition occurs predominantly in the negro; and (2) to devise a method of treatment which will resolve the condition most quickly and safely. This series of 14 cases is the largest to be reported.

Materials and methods

Fourteen consecutive cases of congenital total eversion of the upper eyelids seen by the author during a period of over 3 years (June 1975 to December 1978) at the Eye Clinic, Korle Bu Teaching Hospital, Accra, were studied for: history of the pregnancy and the delivery; occurrence of the condition in siblings; general physical condition; condition of the unwrinkled eyelids in the unilateral cases; and cytogenic analysis of blood cells.

In addition the eyelids of 150 consecutive newborn babies delivered at the Korle Bu Teaching Hospital were examined within 24 hours of birth. The methods of treatment of the 14 cases were as shown in Table 1.

**Table 1 Methods of treatment for the various groups**

<table>
<thead>
<tr>
<th>Group A</th>
<th>Group B</th>
<th>Group C</th>
</tr>
</thead>
<tbody>
<tr>
<td>(a)</td>
<td>(b)</td>
<td></td>
</tr>
<tr>
<td>Lid suture</td>
<td>Subconjunctival hyaluronidase injection</td>
<td>Subconjunctival hyaluronidase injection</td>
</tr>
<tr>
<td>Lid suture</td>
<td>Subconjunctival hyaluronidase injection</td>
<td>Subconjunctival hyaluronidase injection</td>
</tr>
<tr>
<td>Antibiotic ointment</td>
<td>Antibiotic ointment</td>
<td>Frequent instillation of antibiotic ointment</td>
</tr>
<tr>
<td>Eye pad</td>
<td>Eye pad</td>
<td>Eye pad</td>
</tr>
</tbody>
</table>

**Group A. Lid suture only.** A double armed 3/0 silk suture was passed from the upper palpebral conjunctiva near the fornix to the skin just below the eyebrow and tied over a rubber bead. Antibiotic eye ointment was instilled and the eye padded. The eye was dressed daily. The suture was removed when the lid oedema and chemosis had settled well that recurrence of the eversion seemed unlikely.

**Group B. Subconjunctival hyaluronidase injection and lid suture.** 750–1500 units of hyaluronidase were injected into the chemotic conjunctiva before the lid suture was placed. Antibiotic eye ointment was instilled and the eye padded.

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Group C. (a) Subconjunctival injection of hyaluronidase and frequent instillation of antibiotic eye ointment for one eyelid; (b) lid suture after subconjunctival hyaluronidase injection, antibiotic eye ointment, and padding for the other eyelid.

Only topical anaesthesia with amethocaine HCl eye drops was used in all the cases. The first case in the study was allotted to treatment group A by ballot. The subsequent cases were allotted to groups A and B alternately. After 12 cases had been so treated it was decided that subsequent bilateral cases would be treated with subconjunctival hyaluronidase injection followed by lid suture for the worse eyelid and only hyaluronidase injection for the other eyelid (group C).

Results

Deliveries were spontaneous and not prolonged in 13 cases. In case 13 delivery was by caesarean section because of prolonged labour due to cephalopelvic disproportion. None gave a history of previous occurrence of the condition in a sibling or any other member of the extended family. General physical examination showed normal healthy newborn babies in all cases. In the unilateral cases the unaffected upper eyelid was invariably found to overlap the lower eyelid margin partially or totally (Fig. 1). Cytogenic analysis showed no case of Down's syndrome or any other abnormality (Table 2).

Of the 150 consecutive newborn babies whose eyelids were examined 22 showed upper eyelids which overlapped the lower eyelid margins in varying degrees. In 11 of these babies the upper eyelid overlapped the temporal one-third of the lower eyelid margin. In 8 babies the upper eyelid overlapped the temporal half of the lower eyelid margin. In the remaining 3 babies (i.e., 6 eyes) the whole of the lower eyelid margin was overlapped by the upper eyelid.

Fig. 2 shows the length of time it took for the upper eyelids to revert to normal position in each of the cases. In group A the longest time was 4 days. In case 7 of group A the suture was removed on the second postoperative day, and although the eyelid was reverted there was still some oedema, and the eyelid everted again when the baby cried. The eyelid was reverted and padded for 1 day, after which there was no recurrence of the eversion.

In group B the eyelids were reverted with little or no oedema on the first postoperative day. The suture could be removed on the first or the second postoperative day without recurrence of eversion of the eyelid.

Table 2  Findings in the 14 cases of congenital eversion of the upper eyelids

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age/sex</th>
<th>Delivery</th>
<th>Laterality of eversion</th>
<th>General physical condition</th>
<th>Cytogenetic analysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3 days, male</td>
<td>Normal</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>2</td>
<td>3 days, male</td>
<td>Normal</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>3</td>
<td>14 h, male</td>
<td>Normal</td>
<td>Unilateral (left)</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>4</td>
<td>2 days, female</td>
<td>Normal</td>
<td>Unilateral (left)</td>
<td>Normal</td>
<td>Normal female</td>
</tr>
<tr>
<td>5</td>
<td>1 day, female</td>
<td>Normal</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal female</td>
</tr>
<tr>
<td>6</td>
<td>1 day, male</td>
<td>Normal</td>
<td>Unilateral (right)</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>7</td>
<td>1 day, male</td>
<td>Normal</td>
<td>Unilateral (right)</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>8</td>
<td>1 day, male</td>
<td>Normal</td>
<td>Unilateral (right)</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>9</td>
<td>1 day, male</td>
<td>Normal</td>
<td>Unilateral (left)</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>10</td>
<td>6 h, female</td>
<td>Normal</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal female</td>
</tr>
<tr>
<td>11</td>
<td>3 days, female</td>
<td>Normal</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal female</td>
</tr>
<tr>
<td>12</td>
<td>2 days, male</td>
<td>Normal</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>13</td>
<td>1 h, male</td>
<td>Caesarean section</td>
<td>Bilateral (left &gt; right)</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
<tr>
<td>14</td>
<td>1 day, male</td>
<td>Normal</td>
<td>Bilateral (right &gt; left)</td>
<td>Normal</td>
<td>Normal male</td>
</tr>
</tbody>
</table>
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In group C, where conservative treatment with only subconjunctival injection of hyaluronidase was compared with lid suture after hyaluronidase injection, the eyelids with lid suture were reverted with only little oedema on the first postoperative day (treatment b) (Figs. 3 and 4), whereas the eyelids without lid suture reverted on the eighth day in 1 case and on the ninth in the other (treatment a).

Discussion

LATERALITY
The 6 cases of unilateral congenital total eversion of the upper eyelid among the 14 cases in this series suggest that unilateral occurrence may not be rare.

AETIOLOGY
As noted above, Duke-Elder suggested that the eversion of the eyelids is brought about mechanically as the head traverses the birth canal. Stillerman et al. suggested that facial trauma against the uterus during delivery may cause interference with the venous drainage in the eyelids and peri orbital tissues, resulting in the eversion. Gershanik et al. suggested that the normal uterine contractions may provoke the eversion by fetal internal rotation and extension.

In view of (a) the rarity of the condition, (b) its predominance in negro babies, (c) the occurrence of most cases after normal labour and delivery, it is likely that there might be a predisposing factor inherent in the eyelids of these babies. Histological examination of such an eyelid showed no abnormality. On clinical examination of the eyelids of 150 newborn babies at Korle Bu Teaching Hospital the only significant finding was the overlapping of the lower eyelid margin by the upper eyelid in varying degrees in 22 of the babies. This was thought to

Fig. 2. Histogram showing day of permanent reversion of the eyelid.

Fig. 3. Case 13, bilateral congenital total eversion of the upper eyelids before treatment.

Fig. 4. Case 13 on first postoperative day. The left upper eyelid is reverted with little oedema, but the right is still evverted with gross chemosis of the conjunctiva. Both upper eyelids had hyaluronidase injections, but the left one had a lid suture in addition.
be all the more significant as the unverted upper eyelid in the unilateral cases had been found to overlap the lower eyelid margin in all the 6 cases. It is a common experience that pushing the lower eyelid under the upper eyelid makes it easy to evert the upper eyelid even with one hand. It is conceivable that given the condition of a considerable overlapping of the lower eyelid margin by the upper eyelid, and the face in a suitable position, the normal uterine contractions may cause eversion of the upper eyelid. Irritation of the exposed conjunctiva may provoke acute blepharospasm which prevents spontaneous reversion. Trauma to the conjunctiva may then lead to chemosis, which further prevents reversion of the eyelid. The unilateral cases may be due to asymmetrical overlapping of the eyelids. Unfortunately there was no opportunity to examine a comparable number of white newborn babies to see if this presumed predisposing factor was lacking in them. The fact that case 13 was delivered by caesarean section suggests that passage through the whole of the birth canal is not an essential part of the mechanism of eversion.

TREATMENT

Different methods of treatment of congenital eversion of the upper eyelids have been suggested. It seems that the most commonly practised is a purely conservative method whereby antibiotic ointment is applied to the eyes frequently to avoid infection and drying of the conjunctiva.8 10 11 15 Stern et al.7 covered the eyes with clear plastic food wrap to prevent desiccation. This method of treatment has led to reversion of the eyelids in 2 to 3 weeks. Abiose8 mentions the use of subconjunctival injection of hyaluronic acid (presumably hyaluronidase) in the unilateral case she reported, where reversion occurred in 6 days. Surgical methods of treatment have included tarsorrhaphy12 and excision of redundant conjunctiva (quoted by Stillerman et al.13).

In group A in this study, in which only lid sutures were placed, the eyelids reverted permanently in 4 days at most. The eyelids were in their normal position without overlapping within 1 month. In group B, in which subconjunctival injection of hyaluronidase preceded the placement of the lid sutures, the eyelids were reverted with little or no oedema the following day. The sutures could be removed on the first or second postoperative day and the eyelids remained reverted. Overlapping of the lower eyelid margin by the upper eyelid did not persist beyond 2 weeks. In the case reported by Stern et al.7 the eyelids were still overlapping in the published picture taken 2 months after birth.

At this stage in the study it became obvious that subconjunctival hyaluronidase injection followed by the lid suture leads to permanent reversion of the eyelid in a much shorter time than the use of the lid suture alone. It was then decided that the effect of hyaluronidase injection alone be compared with that of the injection followed by the lid suture. In the subsequent 2 bilateral cases the worse eye was treated with subconjunctival hyaluronidase injection followed by the lid suture and the other eyelid with the subconjunctival injection alone (group C). Although there were only 2 cases in this group, the results showed conclusively the superiority of the method used for group B over the conservative treatment with hyaluronidase injection and application of ointment alone. The more oedematous eyelids which had lid suture and hyaluronidase injection were reverted with only little oedema the next day, whereas the conservatively treated eyelids took 8 and 9 days respectively to revert.

Conclusions

A method of treatment which resolves this eyelid condition in one or two days is of advantage as it reduces the chances of desiccation and infection of the exposed conjunctiva. It also reduces the length of stay in hospital. Subconjunctival injection of hyaluronidase followed by passing the lid suture was found to be safe and led to rapid reversion of the everted eyelid. This method of treatment is indicated in cases of congenital total eversion of the upper eyelid with severe chemosis.
Addendum. A day-old Liberian baby with collodion skin disease and bilateral congenital total eversion of the upper eyelids was seen by the author at the Firestone Plantation Company Medical Centre in Harbel, Liberia, in July 1978 (Fig. 5). A similar case was reported by Shapiro et al. in 1969.) Delivery was outside the hospital but was alleged to have been normal. This case was not included in the study.

I thank Dr Edwin Jallah, the Medical Director of the Firestone Plantation Medical Centre, Harbel, Liberia, for permission to publish the picture of the collodion baby. I am also grateful to colleagues who referred cases to me, to Miss Emelia Addo who took the pictures, and to Mr Charles Vimenyo for secretarial assistance.

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