The child

CASE

of
crocodile

It

abducens
described
in
The
Kirkham,9
in
to
from
an
eye
musculofascial anomaly, but
and
eye
and
rare.
A
the
from
an
main feature is
of
condition
of
British
Journal
cases
bilateral
an
associated
on
movement
of
electromyography3-7
of
the
abducens
disorders.
Duane's
syndrome,6
was
limited
in
each
eye
with
retraction
and
upshoot
of
the
adducting
eye,
and
in
addition
adduction
and
convergence
were
slightly
reduced.
Investigation
of
the
lacrimal
defect
confirmed
abundant
lacrimation
on
chewing
a
sweet.
This
was
also
present
on
chewing
a
tasteless
rubber.
Although
nasal
sensation
was
normal,
no
lacrimation
was
produced
on
stimulating
the
nose
with
a
cotton
wool
wisp
or
ammonia
fumes.
General
examination
revealed
a
blurred
expression
but
normal
emotional
and
voluntary
expression.
The
distal
panax
of
the
ring
finger
of
each
hand
was
markedly
deviated
in
the
ring
finger.
X-rays
of
skull
and
chest
were
normal.
Eye
movement
recordings
were
carried
out
by
the
Medical
Research
Council's
Hearing
and
Balance
Unit
at
the
National
Hospital,
Queen
Square,
London,
revealed
that
horizontal
and
vertical
saccades
could
be
generated
in
each
eye
in
response
to
vestibular
and
optokinetic
stimuli.
Vestibulospinal
reflexes
were
normal
as
judged
by
free-fall
electromyography.11
Stapedial
reflexes
were
normal,
indicating
a
normal
innervation
of
the
stapedius
muscle
by
the
facial
nerve.
The
5-tone
audiogram
was
normal.

SUMMARY

The
occurrence
from
birth
of
copious
lacrimation
on
eating
in
some
patients
with
Duane's
syndrome
suggests
that
both
are
caused
by
dysgenesis
or
a
lesion
in
the
vicinity
of
the
abducens
nucleus
in
the
pons.

The
legend
that
crocodiles
wept
before
devouring
their
victims
gave
birth
to
the
term
'crocodile
tears' (gustolacrical
reflex)
which
is
now
used
for
the
condition
of
copious
lacrimation
associated
with
eating.
Reports
of
acquired
crocodile
tears
frequently
appear,1
the
symptom
usually
following
traumatic
or
inflammatory
conditions
of
the
facial
or
greater
superficial
petrosal
nerves.
It
is
widely
believed
that
the
paroxysmal
lacrimation
results
from
the
misdirection
of
regenerating
parasympa-
thetic
secretomotor
fibres
subserving
salivation.
Reports
of
cases
of
congenital
crocodile
tears
are
rare.
A
summary
of
all
reported
cases
(Table
1)
shows
that
in
all
but
the
case
of
Antonelli,
there
is
an
associated
abducens
palsy
or
Duane's
syndrome,
and
that
in
unilateral
cases
the
lacrimal
abnormality
and
eye
movement
disorder
are
ipsilateral,
while
in
bilateral
cases
the
movement
disorder
is
also
bilateral.

Duane's
syndrome2
was
originally
described
as
a
musculofacial
anomaly,
but
more
recent
studies
based
on
electromyography8-7
suggest
that
the
main
feature
is
co-contraction
of
the
horizontal
eye
muscles,
which
distinguishes
Duane's
syndrome
from
an
abducens
palsy.
Retraction
is
often
incon-
spicuous,
and
Huber
et
al.8
pointed
out
that
coc-
contraction
may
be
revealed
only
by
electro-
myography.
It
is
possible
that
cases
described
as
having
an
abducens
palsy
have
in
reality
a
Duane's
syndrome.
The
diverse
associated
anomalies
to
in
Table
1
follow
closely
the
pattern
of
anomalies
described
in
Duane's
syndrome
by
Sachsenweger,3
Kirkham,9
and
Pfaffenbach
et
al.10
The
occurrence
of
crocodile
tears
sheds
some
light
on
the
causative
lesion
in
Duane's
syndrome.

Case
reports

CASE

1

The
child
was
a
boy
aged
3
years
4
months
who
was
the
product
of
a
normal
pregnancy.
Shortly
after
birth
the
parents
had
noted
that
there
was
no
lacrimation
from
either
eye
when
the
child
cried.
From
the
time
when
solid
foods
were
first
taken,

Correspondence
to
Dr
D.
Taylor.

518
Congenital crocodile tears: a key to the aetiology of Duane’s syndrome

Table 1  Summary of all reported cases

<table>
<thead>
<tr>
<th>Case</th>
<th>Crocodile tearing</th>
<th>Emotional tearing</th>
<th>Facial weakness</th>
<th>Duane’s syndrome or sixth nerve palsy</th>
<th>Other features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antonelli55</td>
<td>Unilateral (R)</td>
<td>Absent</td>
<td></td>
<td>Uni/bilateral</td>
<td></td>
</tr>
<tr>
<td>Lutman†</td>
<td>1 Unilateral (?)R/L</td>
<td>• • •</td>
<td></td>
<td>Uni/bilateral</td>
<td>•</td>
</tr>
<tr>
<td>Lillie37</td>
<td>2 Bilateral</td>
<td>• • •</td>
<td></td>
<td>Uni/bilateral</td>
<td>•</td>
</tr>
<tr>
<td>D’Ermo3'</td>
<td>3 Bilateral</td>
<td>• • •</td>
<td></td>
<td>Uni/bilateral</td>
<td>•</td>
</tr>
<tr>
<td>Cricchi38</td>
<td>? Uni/bilateral</td>
<td>• • •</td>
<td></td>
<td>? Uni/bilateral</td>
<td>Oxyccephaly, facial asymmetry</td>
</tr>
<tr>
<td>Jampel, Titone8</td>
<td>Bilateral</td>
<td>Absent</td>
<td>Absent</td>
<td>Bilateral Duane’s</td>
<td>Hypochromia iridis, facial asymmetry</td>
</tr>
<tr>
<td>Sarda et al.41</td>
<td>Unilateral (R)</td>
<td>• Absent</td>
<td></td>
<td>Unilateral palsy (R)</td>
<td></td>
</tr>
<tr>
<td>Regenbogen, Stein42</td>
<td>Unilateral (L)</td>
<td>Present</td>
<td>Absent</td>
<td>Uni/bilateral Duane’s (R)</td>
<td>EMG features of Duane’s syndrome</td>
</tr>
<tr>
<td>Uemura, Tamura45</td>
<td>1 Bilateral</td>
<td>Absent</td>
<td>Absent</td>
<td>Bilateral Duane’s</td>
<td>Bilateral deafness</td>
</tr>
<tr>
<td>Lillie37</td>
<td>2 Bilateral</td>
<td>Absent</td>
<td>Absent</td>
<td>Bilateral Duane’s</td>
<td>EMG features of Duanes’s syndrome</td>
</tr>
<tr>
<td>Cricchi38</td>
<td>3 Bilateral</td>
<td>Absent</td>
<td>Absent</td>
<td>Bilateral Duane’s</td>
<td>Supernumary auricle</td>
</tr>
<tr>
<td>Jampel, Titone8</td>
<td>? Uni/bilateral</td>
<td>• Absent</td>
<td></td>
<td>? Uni/bilateral</td>
<td></td>
</tr>
<tr>
<td>Brik, Athayde34</td>
<td>Bilateral</td>
<td>• Absent</td>
<td></td>
<td>Bilateral Duane’s</td>
<td>Klippel-Feil anomaly</td>
</tr>
<tr>
<td>Trieschmann50</td>
<td>1 Bilateral</td>
<td>Absent</td>
<td></td>
<td>Uni/bilateral</td>
<td></td>
</tr>
<tr>
<td>Lillie37</td>
<td>2 Bilateral</td>
<td>Absent</td>
<td></td>
<td>Uni/bilateral Duane’s (R &gt; L)</td>
<td>Bilateral deafness, deformity of auricles</td>
</tr>
<tr>
<td>Ramsay, Taylor</td>
<td>1 Bilateral</td>
<td>Absent</td>
<td>Absent</td>
<td>Bilateral Duane’s</td>
<td>Deformity of fingers</td>
</tr>
<tr>
<td>Lillie37</td>
<td>2 Bilateral</td>
<td>Absent</td>
<td>Absent</td>
<td>Bilateral Duane’s</td>
<td></td>
</tr>
</tbody>
</table>

*Indicates that the presence or absence of the feature was not recorded.

Case 2

The child, a boy aged 3 years 8 months when first seen, was the product of a full-term normal pregnancy. The parents had noted that from birth copious lacrimation occurred from both eyes on breast feeding. It continued after the child started taking solid foods. When eating dry foods he drank repeatedly. There had been no lacrimation from either eye when crying, and the parents were unaware of any eye movement disorder. He had been deaf from birth. There was no relevant family history or past medical history. On examination the vision was 3/6 in each eye by the Ffooks symbols. There was a small left, alternating convergent squint and a bilateral Duane’s syndrome with marked limitation of abduction and slight retraction on attempted adduction in each eye. Adduction and convergence were reduced. Investigation of the lacrimal defect showed lacrimation on chewing a sweet or a tasteless rubber. Corneal sensation was normal, but no lacrimation was produced on stimulation of the cornea with a cotton-wool wisp or ammonia fumes.

General examination revealed a bland facial expression but normal emotional facial movements. Examination of the ears revealed a rudimentary right auricle and atresia of the right external auditory meatus. The tympanic membrane could not be seen. The left auricle and external auditory meatus and drum appeared normal. There was response only to the loudest auditory stimuli, and it was felt that even this slight response might be to noise-induced vibration of the skull. There was a marked bilateral pes planus.

X-rays of skull, spine, and chest appeared normal. Eye movement recordings revealed a complete inability to generate horizontal saccades in either eye in response to vestibular or optokinetic stimuli. Small visually evoked vertical saccades could be generated but there was no vertical doll’s head response. Vestibulospinal reflexes were absent on free-fall electromyography.

In these cases the features of bilateral abduction weakness and bland facial expression are suggestive of the Moebius syndrome. In case 2 the additional feature of a marked disturbance of vestibular mediated reflexes may be suggestive of a widespread lower brain stem disturbance.

Discussion

Two main theories have been advanced to account for the electromyographic feature of Duane’s syndrome. One theory supported by Blodi et al.5
and Sato\(^9\) suggests that the defect lies at a supranuclear level, while the other supported by Hoyt and Nachtigäller\(^{12}\) and Huber\(^{18}\) proposes that the defect is peripheral, there being a developmentally defective innervation of the lateral rectus by the abducens nerve with a variable pattern of innervation of the other ocular muscles.

The existence of a supranuclear pontine centre for lateral gaze was postulated by Crosby.\(^{14}\) Carpenter \textit{et al.}\(^{15}\) suggested that in monkeys this centre is near or within the abducens nucleus. They showed that unilateral stereotactic destruction of the abducens nucleus was followed by gaze deviation to the opposite side. In the tissue studies of the same animals the preterminal degeneration was most marked in the contralateral medial longitudinal fasciculus and distributed preferentially to the ventral nucleus of the contralateral oculomotor nuclear complex (cells whose axons innervate the contralateral medial rectus). Similarly, Graybiel and Hartweig\(^{16}\) showed that \(^{3}H\) proline injected into the abducens nucleus of cats appeared within the contralateral ventral nucleus of the oculomotor nuclear complex. They proposed that these interneurons are the cells essential in conjugate lateral gaze. A theory that seeks to explain Duane's syndrome on the basis of a supranuclear lesion must therefore account for the normal action of the contralateral medial rectus.

The innervational theory suggested by Huber \textit{et al.}\(^{4}\) and expanded by Hoyt and Nachtigäller\(^{12}\) proposed that a maldevelopment of the abducens nucleus occurs, resulting in innervation of the lateral rectus by fibres of the oculomotor nerve, the variable electromyographic features reflecting the innervation of the lateral rectus by different branches of the oculomotor nerve. Unfortunately the anatomical evidence for substitute innervation is inconclusive, since no case of Duane's syndrome has been examined at necropsy with both histological examination of the brain stem and dissection of the nerves as far as the eye muscles.

The post-mortem study of a case of Duane's syndrome by Matteucci\(^{17}\) showed aplasia of the abducens nucleus. A case of bilateral abducens nerve palsy\(^{18}\) showed the absence of both abducens nerves with diminutive nuclei. Some anatomical support for substitute innervation has been drawn\(^{18}\) from cadaver dissections (cases without information regarding eye movements in life) in which absence of the abducens nerve was associated with innervation of the lateral rectus by the oculomotor nerve.\(^{18-22}\)

However, these findings must be interpreted with caution. Svitzer\(^{23}\) in cadaver dissections and Kimmel\(^{24}\) in histological studies of embryo rabbits have shown that 'anastomosis' between the oculomotor nerve and the abducens nerve sometimes occurs within the cavernous sinus and orbit. Absence of the abducens nerve in cadaver dissections must also be interpreted with caution. Kimmel\(^{25}\) and Bremer\(^{26-28}\) showed that in some cases the fibres from the abducens nucleus do not emerge from the brains stem at the classical site. The innervation of the lateral rectus by a branch of the oculomotor nerve in the cadaver dissections of Generali,\(^{29}\) Tillack and Winer,\(^{30}\) Munnicks,\(^{31}\) and Fasebeck\(^{32}\) may reflect only an abnormal route by which abducens motoneurons reach their destination. In Tillack and Winer's case the abducens nucleus was reported as normal, which might be taken to suggest that although the 'abducens nerve' could not be found the motoneurons must have existed.

Bremer\(^{33}\) suggested that absence of the abducens nerve might arise through a delay in the development of the ocular musculature, allowing the fibres of the abducens nerve to pass caudally, attracted by the postotic musculature. These fibres would disappear with the involution of the postotic mesoderm. Pfaffenbach \textit{et al.}\(^{34}\) suggested that a teratogenic stimulus at an early stage of embryogenesis caused Duane's syndrome and its associated anomalies. Of 55 children with thalidomide embryopathy studied by Papst,\(^{37}\) 4 had Duane's syndrome, and of those with eye movement disturbances many had facial pareses and ear deformities. He suggested that these disorders occurred between 34 and 39 days after the mother's last menstrual period. The other abnormalities associated with these cases of Duane's syndrome are comparable to those described by Pfaffenbach \textit{et al.}\(^{18}\) Trieschmann\(^{38}\) reported 3 patients with thalidomide embryopathy who showed crocodile tears, abducens nerve palsy, or Duane's syndrome and facial paresis, and 2 showed deformities of the pinnae of the ears.

If Duane's syndrome is of teratogenic origin the presence of crocodile tears in some cases sheds light on the site of the lesion. The close association of the eye movement disorder with the lacrimal disorder, respecting laterality in the unilateral cases and being bilateral in bilateral cases, suggests that a single teratogenic lesion is responsible for both. While the site of the lacrimal centre within the brain stem is uncertain, the similar embryological origin of lacrimal and salivary glands suggests that the centres controlling both lie in the same area. A centre controlling salivation exists in the lower brain stem, but its exact location is uncertain.

Kohnstamm\(^{29}\) and Yagita and Hayama,\(^{30}\) using the technique of retrograde degeneration, suggested a centre in the lateral reticular formation at the level of the facial nucleus. Using stereotactic...
stimulation Magoun and Beaton\textsuperscript{31} in monkeys and Chatfield\textsuperscript{38} and Wang\textsuperscript{35} in cats identified 2 areas in the lower brain stem that yield largely ipsilateral salivary secretion. They confirmed that salivation followed stimulation in the lateral area demonstrated by Kohstamm\textsuperscript{29} and Yagita and Hayama,\textsuperscript{30} but in addition a similar response was obtained from the dorsal paramedian reticular formation. Stimulating the intervening area of the reticular formation gave a limited response. Different interpretations were put on these findings. Chatfield and Wang thought that the salivary centre lay ventrolaterally near the facial nucleus and that secretion following stimulation in the dorsomedially placed area represented the stimulation of efferent fibres lying within the genu of the root of the facial nerve. Magoun and Beaton,\textsuperscript{31} however, felt that secretion produced by stimulation more laterally in the brain stem reflected the passage of efferent fibres from dorsomedial cells.

In the embryo rabbit Kimmel\textsuperscript{24,31} found a genu only in fibres from cells of the special visceral efferent column that contribute to the motor root of the facial nerve. This suggests that it is incorrect to ascribe secretion from stimulation in the dorsomedial area to stimulation of efferent neurones, and thus the interpretation of Magoun and Beaton\textsuperscript{31} seems more plausible, that is, that the salivary centre lies dorsomedially with efferents passing laterally through the reticular formation. A dorsomedial centre mediating parasympathetic function would not be surprising in view of the position of the Edinger-Westphal nucleus and the dorsal motor nucleus of the vagus. The cells lying rostrally in this dorsomedial column (that is, in closest association with the abducens nucleus) would subserve lacrimal secretion (indeed in the series of Magoun and Beaton 1 animal showed ipsilateral lacrimation on stimulation in this area).

The most logical explanation of the lacrimal and eye movement disorders in Duane's syndrome would be a lesion causing a nuclear degeneration or dysgenesis in the immediate vicinity of the abducens nucleus, the paradoxical aspects of Duane's syndrome and the lacrimal disorder being the result of a substitute innervation of the lateral rectus by fibres from the oculomotor nerve and the lacrimal gland by fibres subserving salivation.

The authors thank Dr Michael Gresty and Dr Michael Halmaggi for carrying out the eye movement recordings on both patients.

References

\textsuperscript{1}Chorobski J. Syndrome of crocodile tears. Arch Neurol Psychiat 1951; 65: 299-318.

\textsuperscript{2}Duane A. Congenital deficiency of abduction associated with impairment of adduction, retraction movements, contraction of the palpebral fissure and oblique movements of the eye. Arch Ophthalmol 1905; 34: 133.


\textsuperscript{13}Huber A. Electrophysiology of the retraction syndromes. Br J Ophthalmol 1974; 58: 293-300.

\textsuperscript{14}Crosby EC. Relation of brain stem centres to normal and abnormal eye movements in the horizontal plane. J Comp Neurol 1953; 99: 437-79.

\textsuperscript{15}Carpenter MB, McMasters RE, Hanna GR. Disturbances of Conjugate horizontal eye movements in the monkey. Arch Neurol 1963; 8: 231-47.


\textsuperscript{17}Matteucci P, I difetti congeniti di abduzione ("congenital abduction deficiency") con particolare riguardo alla patogenesi. Rassegna Italiana d'Ottalalomogia 1946; 15: 345-80.


\textsuperscript{19}Generali G. Considerazioni anatomo-fisiologiche e patologiche intorno il nervo gran simpatico. Ann Universali Med Compilati del Dottore A Omode 1842; 104: 60.


\textsuperscript{21}Manniks. Quoted in Henle J. Handbuch der Systematischen Anatomie des Menschen. Vejweg, 1879.

\textsuperscript{22}Fasebeck. Quoted in Henle J. Handbuch der Systematischen Anatomie des Menschen. Vieweg, 1879.

\textsuperscript{23}Switzer. Quoted in Henle J. Handbuch der Systematischen Anatomie des Menschen. Vejweg, 1879.

\textsuperscript{24}Kimmel DL. Differentiation of the bulbar motor nuclei and the coincident development of associated root fibres in the rabbit. J Comp Neurol 1940; 72: 83-148.


\textsuperscript{29}Kohnstamm O. Der Nucleus salivatorius Chorda tympani (nervi intermedii) Anat Anz 1902; 21: 362-3.

\textsuperscript{30}Yagita K, Hayama S. Uber das Speichelsekretionszentrum.
James Ramsay and David Taylor


Antonelli A. Anomalia funzionale congenita de la glande lacrymale du coté droit. *Clinique Ophthalmologique* 1902; 8: 35-6.


