

Case report

SYNERGISTIC DIVERGENCE : A CASE REPORT

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Abstract Synergistic divergence is a rare congenital syndrome in which the patient has the limitation of adduction with simultaneous abduction when attempting to look in the opposite direction. By electromyographic study, co-contraction of the horizontal recti, as seen in Duane's retraction syndrome, has been reported. This condition may occur as an isolated entity or in combination with other ocular or systemic abnormalities. We report a case of synergistic divergence with a cleft lip, cleft palate and short stature. Interestingly, a vertical component was also present in this case. **Chiang Mai Med Bull 2003;42(1):31-36.**

Keywords : synergistic divergence, Duane's retraction syndrome

Synergistic divergence is a rare condition. This congenital syndrome has the limitation of adduction in the affected eye and simultaneous abduction of attempted lateral gaze on the unaffected side.⁽¹⁾

The first patient was reported by Barany in 1930.⁽²⁾ Wilcox and coworkers,⁽³⁾ revealed the simultaneous innervation in the medial and lateral recti of the affected eye using the electromyographic technique. This finding was similar to that of Duane's retraction syndrome.

In the literature reviewed, synergistic divergence was reported as an isolated abnormality or in conjunction with other

congenital anomalies, *i.e.* jaw-winking phenomenon, Horner syndrome, congenital ocular fibrosis, oculocutaneous albinism, and arthrogryposis multiplex congenita.^(2,4,5) No report documented an association with a cleft lip, cleft palate or short stature.

We report a case of synergistic divergence associated with other congenital anomalies including a cleft lip, cleft palate and short stature.

Case report

A 27-year old man presented with congenitally abnormal movement of the right eye with moderate left face turn

and chin up. On examination, he had short stature (151.5 cm in height), a scar above the upper lip (Fig. 1), cleft palate, and residual muscle weakness and atrophy as a result of poliomyelitis of the right foot. He had a history of multiple operations for the cleft lip and cleft palate, and the foot abnormality was surgically corrected on June 21, 1998. He was also scheduled for further palatoplasty and cheiloplasty after eye muscle surgery. Upon the first ocular examination (March 15, 1999), he had the best corrected visual acuity of 6/9 in each eye with a left face turn of 30 degrees and slight chin elevation. Manifest refraction measured $-0.50 -2.00 \times 110^\circ$ in the right eye and $-0.50 -1.00 \times 90^\circ$ in the left one. The muscle balance

showed right exotropia 30 prism diopters at distance and 40 prism diopters at near by using the Krimsky test with right hypotropia 20 prism diopters at near in a primary position. The deviation angle of right exotropia significantly increased on the left gaze. Ocular rotation showed a limitation of adduction and elevation of the right eye with simultaneous abduction while attempting left gaze. He also had globe retraction on upgaze (Fig. 2). Not only did the right abduction increase on attempted left gaze, but right hypotropia also increased. The Worth-four dot test showed right suppression at distance and alternate suppression at near. The remaining ocular examination was within normal limits. Glasses were prescribed.



Figure 1. Right exotropia and hypotropia with left face turn and chin elevation.

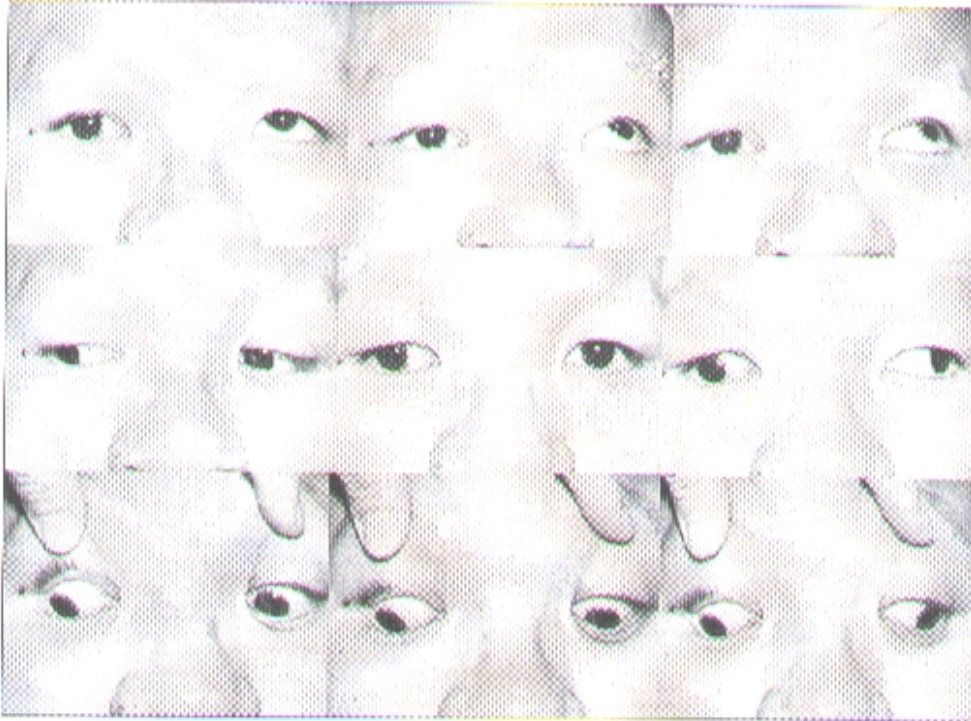


Figure 2. Preoperative ocular rotations showed right exotropia and hypotropia with simultaneous abduction on left gaze

Two months later, the patient was re-examined and the finding was essentially the same. He was happy with his glasses. He came back six months later and requested eye muscle surgery in order to correct the face turn and alignment in the primary position. The muscle balance was re-evaluated, the finding was not significantly changed, and the operation was then scheduled. The patient underwent strabismus surgery under general anesthesia. The forced duction test was performed intraoperatively and revealed positive 3+ restriction of the right medial rectus and 2+ restriction of the right inferior rectus. The lateral rectus muscle

was receded by 9 mm and the inferior rectus muscle by 5 mm in order to correct both exotropia and hypotropia. The medial rectus was not scheduled for resection, due to the presence of preoperative globe retraction.

Eight days postoperative muscle balance the patients had orthotropia in the primary position at both distance and near, however, the simultaneous abduction on attempted left gaze was still present, although in less magnitude (Fig. 3). Two months postoperatively, the muscle balance was right exotropia 14 prism diopters by using the Krimsky test at both distance and near. The last follow



Figure 3. On the first day, postoperative ocular rotations showed orthotropia in primary position.

up at 4 months showed minimal left face turn, and right exotropia 25 prism diopters without vertical deviation, yet the appearance was cosmetically acceptable.

Discussion

Clinical characteristics of a patient with synergistic divergence are congenital adduction palsy, simultaneous abduction on attempted opposite gaze and contralateral face turn to avoid diplopia in the primary position.^(2,6) The abnormalities are usually recognized in childhood and may be unilateral or bilateral.⁽⁷⁾ About 78% of cases affects the left eye.⁽⁸⁾ In the primary position, there is exotropia in the affected eye. In some cases there is also spontaneous abduction nystagmus, and all cases have evoked inverse nystagmus in the affected eye.⁽⁹⁾ Males are slightly more effected than females.⁽⁶⁾

This reported patient had characteristics similar to those previously reported,

however, there were also other abnormalities, including a cleft lip and cleft palate, which were not published in literature.^(2,5)

By electromyographic studies, synergistic divergence is included in the spectrum of Duane's retraction syndrome,^(3,9-11) according to the finding of co-contraction of the horizontal recti, which violates Sherrington's law.⁽⁶⁾

Due to the close association between synergistic divergence and Duane's retraction syndrome, it should be classified as a congenital abnormality, due to the absence or hypoplasia of abducens nucleus,⁽⁸⁾ with the subsequent innervation of lateral rectus muscle by an inferior branch of oculomotor nerve.⁽⁶⁾ The pathogenesis may be the teratogenic disturbance within the second month of pregnancy.^(2,9)

In terms of surgical correction to improve binocularity and/or abnormal

head posture, none of the surgical techniques are the best approach. Most of them include lateral rectus weakening (recession, tenotomy, extirpation), medial rectus resection and various transposition procedures for the medial rectus muscle.^(6,12) The surgical results have varied, but most of the reported cases could not eliminate the simultaneous abduction postoperatively, although the exotropia in the primary position decreased in magnitude. This reported case had both 9-mm lateral rectus recession and 5-mm inferior rectus recession to correct horizontal deviation, vertical deviation and decreased globe retraction in upgaze, which was different from those cases reported in literature. In the short term follow-up, the result was satisfactory.

Conclusion

Aberrant innervation of the oculomotor nerve includes various manifestations. Synergistic divergence is one of those that is rare. Our patient had the congenital limitation of adduction in the right eye with simultaneous abduction on the left gaze. This condition may be the only abnormality or it occurs with other congenital anomalies. This paper reports a case of synergistic divergence associated with a cleft lip, cleft palate and short stature. The patient also had hypotropia and globe retraction. The management for ocular deviation was lateral rectus muscle recession and inferior rectus recession without medial

rectus resection due to the presence of globe retraction.

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รายงานผู้ป่วย SYNERGISTIC DIVERGENCE

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บทคัดย่อ ภาวะ synergistic divergence เป็นกลุ่มอาการผิดปกติแต่กำเนิดที่พบน้อยมาก ผู้ป่วย จะไม่สามารถถลอกตาเข้าในได้ร่วมกับมีตาทั้งสองข้างกลอกออกด้านนอกพร้อมกันเมื่อพยายามที่จะมองเข้าใน จากการตรวจกล้ามเนื้อด้วยคลื่นไฟฟ้าพบว่ามีการหดตัวของกล้ามเนื้อถลอกตาในแนวนอนทั้งสองมัดพร้อมกัน คล้ายกับที่พบในผู้ป่วยกลุ่มอาการ Duane อาจพบความผิดปกติอย่างอื่นของตาหรือระบบอื่นร่วมด้วยได้ รายงานผู้ป่วยนี้ มีภาวะ synergistic divergence ร่วมกับ ปากแหว่ง เพดานโหว่ ตัวเตี้ย นอกจากนี้ยังมีความผิดปกติของกล้ามเนื้อตาในแนวตั้งร่วมด้วย เชียงใหม่เวชสาร 2546;42(1):31-36.

คำสำคัญ : synergistic divergence, กลุ่มอาการ Duane
