

LIMITATION OF GAZE

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Abstract

Limitations of gaze may be congenital or acquired. The Optometrist should differentiate the two and look for associated anomalies. Some neurologic and congenital oculomotor anomalies are discussed.

Limitations of gaze may be encountered by the Optometrists in practice either as an isolated problem or in association with other anomalies. These anomalies may be neural, muscular or traumatic in nature and the Optometrist should be able to either reassure his patients or refer them to the appropriate health professional.

Eye movements may broadly be categorized as outlined in Table 1. Table 2 is a summary of disorders of eye movement mechanisms (1). These disorders may be either congenital or acquired. Often torticollis is associated with these anomalies. It should also be noted that with horizontal anomalies of gaze there is

a face turn; vertical anomalies which produce a rise or fall in chin position; and torsional disturbances cause a head tilt. A patient with a right superior oblique paresis will often have the head turned to the left, face to the right and chin down, (Figure 1).

Acquired Limitations of Gaze

Acquired limitations of gaze are primarily due to trauma such as motor vehicle accidents and often involve orbital fractures. Greenwald, Kenney and Shannon (2) did a retrospective study of 128 patients with orbital fractures. Table 3 summarizes the signs and symptoms of orbital fracture. Table 4 lists complications of surgery to repair orbital fracture which may be encountered by the optometrist (2).

Other causes of acquired limitations of gaze may include internal carotid artery aneurysms affecting oculomotor nuclei in the midbrain region, aberrant third nerve regenera-

tion, viral infections, hydrocephalus, tumors, orbital congestion, ophthalmoplegic migraine, multiple sclerosis, and Myasthenia gravis. These conditions are often associated with other signs and symptoms but are acquired. A development or congenital anomaly can often be differentiated from the acquired type by obtaining early childhood photographs and noting consistent abnormal head positions.

Congenital Limitations of Gaze

Congenital ocular motor paralyses may be due to developmental abnormalities of ocular motor nerves and extraocular muscles (which is to be discussed in this article), developmental abnormalities of the brain, and generally other conditions such as developmental ophthalmoplegias myasthenia congenita and cyclic oculomotor paralysis.

Developmental abnormalities of ocular motor nerves and extraocular muscles are outlined in table 5 (3)



Fig. 1 Right Superior Oblique Paresis

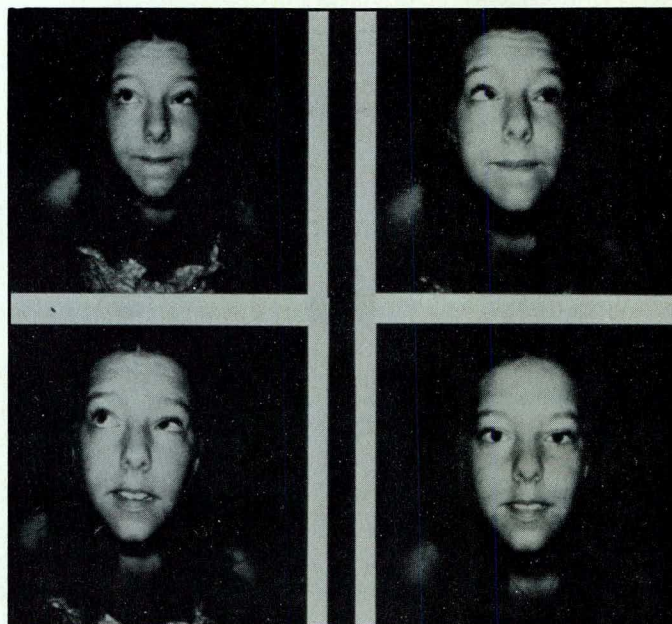


Fig. 2A Brown's Syndrome

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tion, aberrant third nerve regeneration, viral infections, hydrocephalus, tumors, orbital congestion, ophthalmoplegic migraine, multiple

and special examples include Möbius syndrome, Duane's retraction syndrome, Brown's tendon sheath syndrome and strabismus fixus.

Möbius Syndrome

Möbius Syndrome is due to a congenital aplasia of the VI and VII nerve nuclei. There is a facial diplegia, often symmetrical and an inability to abduct the eyes. Associated findings include lingual palsy, external ophthalmoplegia, malformations of extremities and digits, branchial malformations pectoral muscle defects and mental deficiency.

Strabismus Fixus

This is a rare type of congenital esotropia in which the medial rectus muscles are tight and inelastic. Both eyes are convergent. There is difficulty in abducting the eye with forced ductions. The medial recti are replaced by fibrous bands.

Superior Oblique Tendon Sheath Syndrome (Brown)

Brown first described this structural anomaly in 1950. There is shortening of the anterior portion of the superior oblique tendon sheath which restricts passive elevation in the primary position and abduction (Fig. 2 and 3). There is an absence of or minimal overaction of the ipsilateral superior oblique muscle. Crawford reported on the surgical treatment of 28 patients with Brown's syndrome and states that "My results (as well as other surgeons) with similar surgery have been disappointing". (4) Since this anomaly does not interfere with binocular vision in the primary position of gaze not in reading it is best left untreated.

Duane's Retraction Syndrome

Duane first described this syndrome in 1905 which produces six characteristic anomalies of the affected eye: It consists of

1. Decreased abduction
2. Decreased adduction
3. Retraction into the orbit on adduction
4. Oblique elevation or depression on adduction
5. Partial closure of the eyelids on adduction.
6. Deficient convergence



Fig. 2B Brown's Syndrome



Fig. 3 Duane's Syndrome

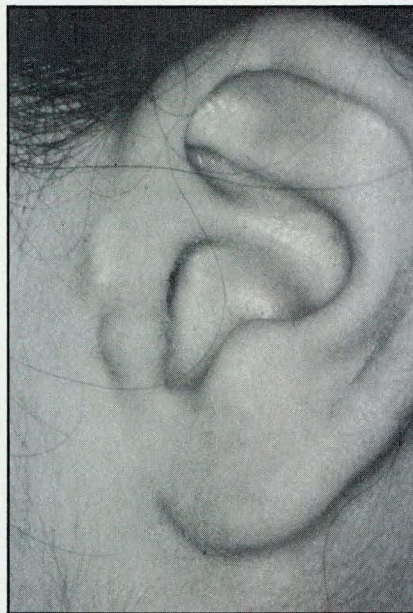


Fig. 4 Wildervanck's Syndrome

(Courtesy Dr. T. Kirkham)

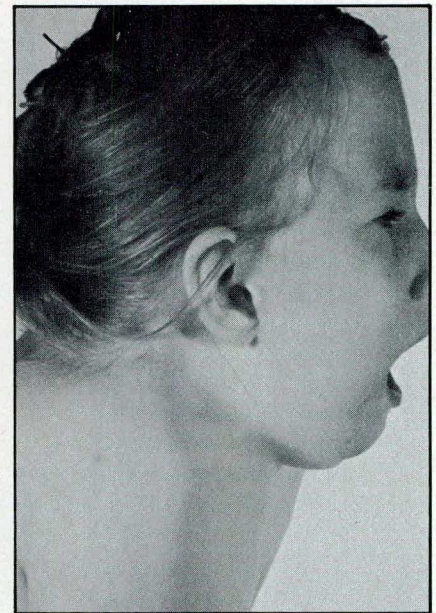


Fig. 5 Goldenhar's Syndrome

Clark and Green reported on a clinical-pathologic case of bilateral Duane's Retraction Syndrome (DRS). (5) An autopsy study revealed on absence of the abducens (VI) nuclei and nerves from the brainstem. The lateral rectus muscles were partially innervated by branches from the oculomotor nerves. Approximately 30-50% of patients with DRS have associated congenital defects involving the eye, skeleton, ears and other nerves.

Cross and Pfaffenbach have estimated that the differentiation of these frequently affected structures occurs between the fourth and the eighth week of gestation, and that this coincides with the development of the third, fourth, and sixth cranial nerves. (6) These authors have postulated that a teratogenic event during the second month of gestation could result in DRS. Kirkham reported on a series of 126 patients and concluded that the triad of Duane's

syndrome, the Klippel-Feil anomaly (Congenital strabismus, bilateral DRS, congenital fusion of cervical vertebrae producing a short immobile neck, often with torticollis; spastic paraplegia, deafness and mental deficiency), and perceptive deafness is inherited as a dominant condition with variable penetrance and expression. (7) (Figure 3).

Wildervanck (1952, 1960) described a syndrome with Cervico-Oculo-Acoustic manifestations. The triad consists of a bilateral DRS, and congenital ipsilateral aplasia of the auditory (VIII) and facial (VII) nerve. This syndrome is often associated with the Klippel-Feil Syndrome and may be x-linked dominant. (Figure 4).

Goldenhar's Syndrome (Oculo-auriculo-vertebral dysplasia) consists of: an epibulbar limbal dermoid and coloboma of the lid; Low set ears or ear deformity (which may be associated with deafness) Prewriauricular cutaneous appendages (skin tags); mandibular hypoplasia and occipitalization of atlas hemivertebrae may also be present (Figure 5). The optometrist should note limitation of head motion, short neck, other facial deformities, hearing problems and anomalies of the external ear, as they may be associated with DRS.

Isenberg and Urist, in their study of 101 patients with DRS found a higher incidence in females 58 to males 43 (8). Fifty-six patients presented with a left DRS and 29 a right DRS. Fifteen per cent had a bilateral DRS usually associated with an A-pattern; 36 patients were orthophoric patients, three had head turn, and 2 had hyperphorias. All strabismus patients displayed a head turn with the eyes deviated toward the field of action of the under-acting muscle; 52% demonstrated changes in both eyelids; 18% dropped the upper lid; 21% elevated the lower lid and 7% showed no change. Sixty-seven per cent were hyperopic, 18% myopic and 16% emmetropic; 10% had amblyopia and 50% anisometropia. Kirkham, in a study of 110 patients with DRS, re-

Table 1. Control mechanism (1)

	Position maintenance	Pursuit
Function	Maintain eye position vis-a-vis target	Maintain object of regard near fovea, match eye and target
Stimulus	Visual interest and attention	Moving object near fovea
Latency (From stimulus to onset of eye movement)		125 msec
Velocity	Both rapid (flick, microsaccades) and slow (drifts)	To 100 deg/sec with central scotoma (accurately to 30 deg/sec)
Feedback	—	Continuous

Table 2. Summary of disorders of the eye movement mechanism (1)

Area of lesion	Mechanism involved	Defect
Frontal (a) Unilateral	Saccadic	Absence of contralateral rapid eye movements (refixation saccades, fast phase of vestibular nystagmus, fast phase of OKN); temporary tonic deviation to side of lesion; gaze paretic nystagmus on gaze to side opposite lesion.
(b) Bilateral		Bilateral defects same as above, and absence of vertical saccades
Parietal	Disconnection of occipito-frontal fibers	Defective fast phase of OKN; spasticity of conjugate gaze; cogwheeling, saccadic pursuit
Occipitoparietal (a) Unilateral	Pursuit, vergence position maintenance	Inability to fixate, inability to follow to contralateral side
(b) Bilateral	Pursuit, vergence position maintenance	Cortical blindness, defective vertical following, bilateral horizontal deficits, impersistence of gaze
Capsule/sub-thalamus Pretectum	Saccadic or following Vertical saccadic, pursuit	Defective saccades or following or both Limitation of vertical gaze, retraction nystagmus, pupillary abnormalities, Parinaud's syndrome
Paramedian pontine reticular formation (PPRF)	Saccadic	Absence of ipsilateral saccades, fast phase of vestibular nystagmus, fast phase of OKN
Internuclear ophthalmoplegia (INO)	Saccadic, Pursuit	Abducting nystagmus; supranuclear paresis of ipsilateral medial rectus, sparing convergence
Pontine gaze center	Saccadic, pursuit vestibular	Loss of ipsilateral eye movements
Diffuse supranuclear degeneration (progressive supranuclear palsy, Huntington's chorea)	Saccadic	Loss of saccades, which are replaced by slow eye movements (following?)
Nuclear lesions	All	Loss of all movements in field of muscles controlled by involved subnuclei
Cerebellum	Saccadic, pursuit	Dysmetria, flutter, opsoclonus; saccadic pursuit (cogwheeling)

Table 3
Signs and Symptoms of Orbital Fracture

	No. of Patients (%)
Limitation of extraocular movements	98 (77)
In up-and downgaze	36 (28)
In upgaze	46 (36)
In downgaze	9 (7)
Diplopia	89 (70)
In central upgaze	48 (38)
In central downgaze	29 (23)
Entrapment of the extraocular tissues	68 (53)
Inferior rectus muscle	23 (18)
Inferior Oblique muscle	2 (2)
Orbital tissues (not specified)	48 (38)
Clouding of the maxillary antrum	45 (35)
One side	44 (34)
Both sides	1 (1)
Preoperative hypesthesia of the skin in the distribution of the infraorbital nerve	38 (30)
Forced duction test	
Positive	36 (28)
Negative	7 (5)
Enophthalmos	29 (23)
One side	28 (22)
Both sides	1 (1)
Orbital Emphysema	15 (12)
Retrobulbar hemorrhage	2 (2)

ported that 44 had anisometropia of more than 1 diopter and 23 of these patients were amblyopic. (9)

Surgery is not usually recommended for DRS. The Optometrist should reassure the patient and their family and advise the patient with a unilateral DRS to sit on the side of a classroom where the field of gaze is normal and no diplopia or extreme

TABLE 4
Postoperative Complications or Persistence of Signs and Symptoms

No. of	Patients (%)
Persistent diplopia	21 (20)
Not specified	10
In upgaze	8
In downgaze	5
In abduction	3
In primary gaze	1
In adduction	1
Persistent enophthalmos	11 (11)
Persistent blepharoptosis	2 (2)
Epiphora	1 (1)
Reoperation for diplopia	5 (5)
Vertical muscle surgery	4
Horizontal muscle surgery	1
Reoperation to remove an implant	2 (2)
Extrusion	1
Diplopia	1
Reoperation for epiphora-dacryo-cystorhinostomy, débridement of scar, and medial canthoplasty	1 (1)
Persistent anesthesia of the infra-orbital nerve	1 (1)

head turn will disturb them.

The purpose of this article is to alert the optometrist to the problems and anomalies associated with oculomotor limitations of gaze and to discuss some limitations of treatment. The author wishes to thank Drs. T. Kirkham, J. Little, D. Anderson and J. Wise for the opportunity of seeing their patients at McGill University department of Ophthalmology, Montreal Childrens Hospital, Montreal General Hospital, the Royal Victoria Hospital and Montreal Neurologic Institute over the past five years.

TABLE 5
Congenital Ocular Motor Paralysis (3)

Developmental abnormalities of ocular motor nerves and extraocular muscle:
Absence or hypoplasia of cranial nerve nuclei or nerve fibers.
Aberrant innervation
Dysgenesis (absence or hypoplasia) of extraocular muscle
Abnormal muscle insertion.
Fibrous substitution of extraocular muscle.
Fascial defects (sheath and check ligament abnormalities).

Special examples:
Möbius syndrome.
Duane's retraction syndrome
Brown's tendon sheath syndrome
Strabismus fixus.

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