Scientific Essay:

Describing Duane's (Retraction Syndrome)

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INTRODUCTION

There is now a broad consensus that the traditional classification of Duane's Retraction Syndrome (DRS) should be abandoned. The old "Type I" - eye moves in, not out; Type II - eye moves out, not in; Type III - eye moves neither in nor out" is grossly inadequate for choosing appropriate surgical therapy to address the patient's primary needs and for understanding the etiology of an individual patient's unique pattern of strabismus.

When M. Edward Wilson, MD became chairman of the committee producing Section 6 of the Basic and Clinical Science Course ("Resident's Home Study Course") of the American Academy of Ophthalmology, the volume entitled *Pediatric Ophthalmology and Strabismus*, he radically changed the entry on Duane's to Duane Syndrome and eliminated the Types from the index and, except for a brief historical mention, from the text as well. (Compare references 1 and 2.)

Whenever strabologists gather and talk about patients with Duane's, they generally avoid the Types. Imagine Ed Buckley, MD, moderating the Difficult Problems Strabismus workshop at the annual meeting of the AAPOS saying: "This patient presented with eso-Duane's of the left eye with a large left face turn and few signs of co-contraction". Immediately you understand the primary problem and can think of reasonable surgical alternatives. Wouldn't it be even better if we usually specified a group of characteristics in a routine order, one that is natural to our ears as experienced strabologists?

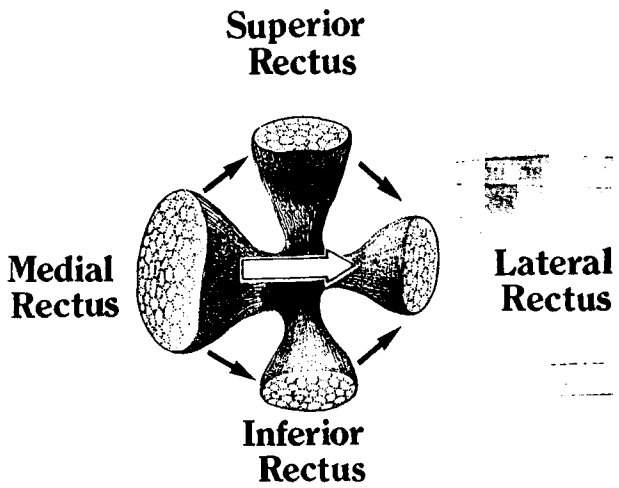

Figure (Mims III): The innervation transfer diagram. The arrows in the diagram show all potential migrations; only one, two, or at most three arrows would serve to explain most patients.

THE INNERVATION TRANSFER DIAGRAM

Please see the Figure. I first presented this diagram in 1983 in a talk at the first Vancouver meeting of the AAPOS. Prior to now, it was only published in a 47 page booklet (3) I had printed at the local Speedy® print and distributed to 50 worthy strabologists at the meeting. This booklet is referenced as reference #121 in Arthur Jampolsky's awesomely comprehensive reference list at the end of his chapter on Duane Syndrome, Chapter 24 in *Clinical Strabismus Management* edited by Arthur L. Rosenbaum, MD and Alvina Pauline Santiago, MD (4).

Also, at the 1996 Snowbird meeting of the AAPOS, Robert S. Baker, MD in a workshop referred to this diagram and pointed out that in the 3 to 4 week old embryo these muscles are touching each other at the back of the orbit and no leap across space is required for these innervation transfers (mis-innervations) to take place. Conceptually, however, this diagram had been helpful to me again and again in understanding the patterns of individual patients with Duane Retraction Syndrome (DRS). **Keep in mind that the various small and large arrows show all potential migrations and that only one or two arrows would serve to explain most individual patients.**

In the third week of embryologic life, the posterior (caudal) half of the sixth cranial nerve sprouting from the brain stem travels caudally to innervate structures associated with the branchial clefts, which later atrophy as embryonic development progresses (5). In most cases of DRS, the entire 6th cranial nerve, instead of merely the posterior half of the nerve, is attracted caudally to these structures without specific teratogenic stimulus (6). Later, the entire 6th cranial nerve and its nucleus atrophy, leaving the developing embryo without a 6th cranial nerve. In 95% of DRS patients this is the only initial abnormality, merely a coincidence of the juxtaposition of the 6th cranial nerve and the tissues that it innervated in the phylogeny being recapitulated in the developing embryo. This is why 95% of DRS patients have no other congenital abnormality (7). In about 5%, other lesions in this area of developing brain are present, producing congenital nerve deafness and/or crocodile tears.

The patterns of ocular motility of DRS are the result of developmental adaptations that occur in the embryo as a result of the absence of the 6th cranial nerve in the orbit. The un-innervated lateral rectus (LR) produces a lateral gradient of neurotropic attraction among the extraocular muscles packed together at the back of the orbit (8,9). The most likely responder to this lateral gradient of attraction is fibers that would normally be innervating the medial rectus (MR), fibers from the MR subnucleus of the 3rd cranial

nerve (10,11). This is why in the diagram the largest arrow (white, outlined in black) indicates transfer of innervation from the MR to the LR. In this common pattern, on attempted adduction, both the MR and the LR are simultaneously innervated. This co-contraction prevents full adduction of the globe and makes the palpebral fissure narrow on adduction. If the co-contraction is severe enough, upshoot and downshoot on adduction occur on a mechanical basis, the "flipping" mechanism popularized by Jampolsky (4).

Note that these "shoots" are not usually due to innervation from the vertical rectus muscles. In many cases of upshoot in adduction there is no exotropia in upgaze, thus indicating the superior rectus (SR) has not contributed fibers to the LR; and in many cases of downshoot there is no exotropia in downgaze, thus indicating that the inferior rectus (IR) has not contributed to the LR.

Jampolsky has claimed that no cases of upshoot in adduction are caused by the SR picking up fibers from the MR. He says you can prove this in a patient with DRS and upshoot in adduction of the left eye by bringing the non-Duane's eye into down and right gaze. He says you will never see a hypertropia of the Duane's eye in this circumstance (4).

One should never say never in DRS; Mohan & Saroha (12) have recently published cases which do demonstrate an "innervational" etiology of upshoot and downshoot, with convincing photographs showing a hypertropia of the Duane's eye with the other eye in abduction and very slight downgaze and another case with a hypotropia of the Duane's eye with the other eye in abduction and slight upgaze (their Figures 3 and 2A, respectively). In these rare cases, the innervational upshoot would be explained by the arrow indicating transfer of fibers from the MR to the SR and the innervational downshoot would be explained by the arrow indicating transfer of fibers from the MR to the IR.

Kushner Syndrome, pseudo overaction of the inferior oblique, incidentally, is fully explained by transfer of fibers from the SR to the LR without any transfer of fibers from the MR to the LR. This is why in Kushner Syndrome you see no hypertropia as the eye is brought back and forth across the primary plane and no narrowing of the palpebral fissure on adduction. When the contralateral eye moves up and out, however, the SR subnucleus in the ipsilateral eye innervates (inappropriately) the LR. Kushner Syndrome has the upshoot of Duane's with no MR-LR co-contraction. (I know, Kushner doesn't claim credit for the first publication of this syndrome; Alan Scott, MD published one case several years earlier, and Scott has been collecting a series of patients who have 6th nerve palsies who have better abduction in upgaze whom he is certain have some SR fibers in the LR.) (13-15)

FIVE MAJOR CHARACTERISTICS TO DESCRIBE

In addition to reporting whether the right eye or the left eye has a unilateral DRS (or indicating which eye one is describing in bilateral DRS), there are five important characteristics to describe:

1. Deviation in the primary position, (binocular misalignment) designated as esoDuane's, exoDuane's, or orthoDuane's, (Or, if you prefer to eliminate the possessive grammar form, esoDuane, exoDuane, or orthoDuane) with any deviation smaller than 5ET or 5 XT being categorized as orthoDuane('s).

2. Face turn consequent to the primary position deviation, (abnormal head posture or AHP) such as a 10° face turn, measured with distance fixation, a laser pointer held above the child's head in the sagittal plane, and remembering that at twenty feet, 21 inches (54 cm) to one side equals 5° (or using an orthopedic goniometer or the CROM device).

3. Signs of co-contraction, (i.e. retraction), each with varied severity and most commonly MR-LR co-contraction, potentially including:

a) Narrowing of the palpebral fissure of more than 1.5 mm on adduction with noticeable enophthalmos on adduction;

b) Limitation of adduction indicated by the Urist test of limitation of adduction with the Hirschberg corneal light reflection 1 mm or more inside the limbus on attempted full adduction. [Urist saw a lot of this when he tried up to 10 mm recessions of one MR for ET.];

c) Exotropia of more than 3 XT in contralateral gaze;

d) Remoteness of near point of convergence, i.e. beyond 7 mm;

e) Severe, moderate, mild or absent upshoot and/or downshoot in adduction.

Note: The above 5 signs of co-contraction are a sensible shortening of longer lists previously published (3,8,9), and are nicely summarized in the Methods section our recent paper by Morad, Kraft & Mims III (16), detailing cases of DRS for whom very small resections and moderate recessions were appropriate because of very few signs of co-contraction present preoperatively. When there is significant upshoot in adduction, it is important to indicate whether it is possible to bring the contralateral eye into abduction in slight downgaze and not see a hypertropia of the Duane's eye, in order to distinguish whether the upshoot is the much more common mechanical ("flipping") type or the much rarer innervational type in which the SR has picked up some innervation from the MR.

4. Exotropia in downgaze, possibly with significant face turn in reading downgaze, and/or **exotropia in upgaze,** which is usually of little clinical importance. XT in upgaze indicates transfer from the SR to the LR and XT in downgaze indicates transfer from the IR to the LR.

5. Severity of limitation of abduction.

EXAMPLES OF ADEQUATE VERBAL DESCRIPTIONS

(Again, imagine Buckley saying these words.)

"Before any surgery was done, this patient began as a garden variety vanilla esoDuane with small left face turn and moderate signs of co-contraction, including moderate narrowing of the palpebral fissure on adduction, moderate limitation of adduction on the Urist test, 5 XT on contralateral gaze, normal near point of convergence

(NPC), and very mild upshoot and downshoot seen only in the extremes of gaze on testing of versions and never noticed by the parents, and no exotropia in upgaze or downgaze and the usual marked limitation of abduction which did bother the parents." (In a diagram drawn for this patient, the white arrow would be smaller in size than in the figure, indicating moderate MR-LR transfer of innervation and no other arrows would be present. This patient has one of the many different sets of characteristics that would have been previously lumped together as Type I.)

"I want the panel to give their first choice to eliminate the unsightly upshoot in adduction in this orthoDuane patient with no significant face turn and several signs of co-contraction, including marked upshoot in adduction, severe narrowing of the palpebral fissure on adduction, positive Urist test indicating severe limitation of adduction, large XT in contralateral gaze, moderately remote but functional NPC, no exotropia in up- or downgaze and severe limitation of abduction. This upshoot is thought to be mechanical in origin, because when the contralateral eye is brought into full abduction and slight downgaze there is no hypertropia of the Duane's eye." At this point you know the panel is going to discuss Y-splits, recess-recess, and recess-recess with split lateral transposition of the vertical rectus muscles [which would probably be unwise if there were an XT in upgaze or downgaze] and isn't going to recommend recession of the Duane's SR. Isn't this really much better than simply calling this patient a Type III? The diagram would show a very large white arrow indicating transfer of a large number of fibers from the MR to the LR, with no other arrows present. Previously, it was said that Type III is merely a Type I with more severe co-contraction.

"One of our members wanted to present this very rare patient for advice. This patient is a left eye exoDuane with large right face turn, abundant signs of co-contraction, including narrowing of the palpebral fissure with enophthalmos on adduction, marked limitation of adduction on the Urist corneal reflection test, large XT in contralateral gaze, remote NPC, world champion upshoot in adduction with exotropia in downgaze and a consequent 10° right face turn in reading downgaze with absolutely full abduction of the Duane's eye."

This is the hen's-teeth-rare Type II, about 1% of Duane's patients, whose 6th cranial nerve is present in the orbit but arrived so late in embryological development that the innervation-hungry LR produced a lateral gradient of attraction and picked up innervation from the MR and the IR. The diagram for this patient would include only arrows from the MR to the LR and from the IR to the LR.

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