

Answers to Audience Questions - WSPOS Worldwide Webinar 4: Duane Syndrome - The Hidden Secrets

WWW 4 Panellists



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1. I like using IO in upshoots. What about the panel?

VS: IO surgery has a specific role in the upshoots especially innervational overshoots, and when suspected to have mixed pattern of overshoots. We should look carefully for the presence or absence of inferior oblique overaction clinically and look for excyclotortion on fundus examination. If it is present and the pattern is suggestive of a slow upshoot on attempted adduction, IO recession can be tried and gives good results.

If you are suspecting a mixed pattern of upshoot, possible repeat the traction test for the IO after LR recession with Y split. If it is still present, we can go ahead with it at the same time as primary surgery or we can do it as a staged procedure.

AA: Yes, this is my preferred technique <https://www.ncbi.nlm.nih.gov/pubmed/23672883>

SK: For me, it depends on whether you are dealing with a mechanical upshoot – due to lateral rectus tightness and slippage, or innervational whereby the other third nerve innervated muscles (superior rectus and/or inferior oblique) are co-innervated with the lateral.

Clinically, if there is a gradual updrift into primary position and adduction, and fundus extorsion, this combination suggests it could be an inferior oblique co-innervation, and weakening of the oblique may make sense. If it is an abrupt upshoot on adduction, usually accompanied by worsening retraction, this suggests a mechanical cause and I would not include oblique surgery, but rather consider a weakening of the LR.

Also see my answer to question 48 below.

DH: This makes sense when there is torsion and when the upshoot is innervational.

SD: If there is vertical in primary gaze and worsen on adduction, plus excyclotorsion, I would add IO recession, otherwise no.

RK: Yes, in cases of innervational upshoot with hypertropia. If the vertical deviation is more than 20 PD and there is mixed type of upshoot one could add Y split or SR recession to IO weakening. The decision depends on the following factors: Excyclotorsion, amount of hyper in PP, FDT findings of LR and SR and any intraoperative findings such as muscle heterotropia, additional muscle band etc.

2. Any role of adjustable sutures in DRS?

VS: Although I personally do not do them routinely but possibly it is reasonable to do adjustable suture on the muscle to be recessed as part of bilateral recessions, or along with transposition procedures to avoid over-corrections. There are couple of patients when we had to advance the medial rectus muscle after SRT in early post-operative period, so doing adjustable suture surgery can be useful in this situation.

AA: In selected cases in adults I find it very useful

SK: Agree. Due to unpredictability of the result in a variable dysinnervation condition like DRS.

DH: I think essential. There are so many variations in DS that the response of any one patient to surgery is difficult to predict.

SD: Yes, I tend to do it in adult with Duane as in children it is a bit challenging unless another quick sedation/GA readily available

RK: Not for me, I do not perform adjustable sutures in these cases.

3. Does anyone use delayed adjustables in Duane?

VS: No

AA: No

SK: I do the final adjustments later on the same day as surgery.

DH: Yes. All of my adjustable suture cases can be adjusted up to 1 week later.

See <https://www.youtube.com/watch?v=hbnjv94yudi>

SD: Not for me as I do all my adjustable a few hours after surgery.

RK: Not for me, I do not perform adjustable eye muscle surgeries.

4. Has Steve added small plications to the hummelsheim in Duane?

SK: No – technically, I find it hard to fold over the lateral such a small amount – Remember you want only 3-3.5 mm. Resection is much easier to measure exactly, and avoid excessive tightening.

5. Nowadays, do we have an idea about what causes Duane syndrome? Can it be transferred from parents to children, I mean, is it hereditary?

VS: Majority of the patients with Duane syndrome are sporadic, however, it is a part of C2D2 disorders and occasionally be inherited. Mutations could be on the various genes: DURS1 locus located on chromosome 8(8q13), The responsible gene, *CHN1*, is located on chromosome 2 (2q31), *SALL4* mutations on chromosome 20 (20q13) which causes Duane radial ray syndrome (with dysplasia of radial bone).

AA: It is quite common to be hereditary. In a study we had on 67 patients with Duane syndrome, only 1 patient showed a positive family history (unpublished)

SK: Agree. It can run in families. I have one family with three consecutive generations with DRS.

It is very likely caused by an insult in the first trimester – around 6-8 weeks' gestation – from studies by Marilyn Miller and associates on thalidomide toxicity and ocular manifestations. And it makes sense, as that is exactly when the ocular motor nerves begin to reach the mesoderm that will become the ocular muscles.

DH: About 2% of cases have a known genetic cause. For a detailed discussion of the genetics of Duane syndrome, see <https://www.ncbi.nlm.nih.gov/books/NBK1190/>

KA: About 90% of Duane cases are sporadic, but some autosomal dominant pedigrees have been reported, associated with a mutation on the *CHN1* gene. I have seen some of these familial cases, and all have been Type I ET Duane.

SD: Yes, Duane syndrome is one type of many congenital cranial dysinnervation disorders (CCDD) and it is hereditary.

YM: There is a great paper by Cameron Parsa: [JAMA Ophthalmol. 2013 Apr;131\(4\):439-47. Thromboembolism and congenital malformations: from Duane syndrome to thalidomide embryopathy. Parsa CF¹, Robert MP.](#)

6. I think we should look for secondary innervation effects on the other eye while deciding about surgery in Exotropic Duane; is that right?

VS: Yes, as Dr. Awadein explained in his talk and others explained, we should try and look for the degree of co-contraction and do limited surgery in the affected eye and greater amount of LR recession in the fellow (normal) eye.

AA: Yes, we should.

DH: Agreed. We should always be looking for anomalous innervation and dysinnervation in these cases. Otherwise you can end up with surprises, especially when operating on the contralateral eye.

SD: Yes, indeed! I often add contralateral lateral rectus simultaneous recession and resection to expand field of BSV in my management of type 2 Duane.

RK: Perform more LR recession in the normal eye and less in the Duane eye. This also helps in further, in limiting the abduction limitation due to LR recession in the Duane. However, contralateral recession can accentuate the retraction on adduction in the Duane eye due fixation duress.

7. Can periosteal fixation be undone in the event of a large undercorrection in exotropic Duane?

VS: There are various techniques of doing periosteal fixation, in some of them the muscle is brought about and attached to periosteum after near the lateral canthus. This is possibly reversible; however, I have never done a reversal as of now.

AA: Yes, it is a reversible procedure, though I never had to undo it.

DH: Agree. Theoretically yes, but despite periosteal fixation there is often residual LR function.

SD: Yes, only if you use Ethibond 4, or 5/0 light blue coloured non-absorbable suture as other sutures hard to find out. Technically it is challenging due to scarring!

YM: Yes

RK: Theoretically yes. But that's not needed in Duane or even in third nerve palsy. Orbital factors also contribute to tightness and usually there is residual XT. I have never reversed one.

8. How will downshifting of Y split work? As splitting of LR, is to be splayed across the globe to prevent the slippage of globe under LR?

VS: After Y splitting the Lateral rectus muscle, the 2 ends can be downshifted by equal amount but are still attached about 1 Tendon width apart. This way the forces are no longer at the upper end of the eyeball but shifted down, while the 2 ends of the split lateral rectus muscle are attached far apart from each other to prevent globe slippage. In this way, both the objectives are fulfilled: The Y split corrects the overshoot and downshift of the split LR corrects the associated vertical squint.

SK: I have never offset the (entire) split muscle down. It is already spread out well above and below the horizontal plane.

SD: I found down shift of LR after Y split is unnecessary as you have already displaced the LR somewhat virtually by splitting.

RK: This is a possible/plausible hypothesis which needs to be proved in more number of cases.

9. What about Y splitting in Exo Duane with globe retraction?

VS: In patients with significant i.e. Greater than 2 globe retraction (i.e. about > 50% narrowing of the palpebral fissure height on attempted abduction, and sometimes with grade 2 retraction) additional MR recession is needed and Y split alone does not work.

In these cases, it is recommended to add MR recession by a small amount and increase the LR recession with Y split (i.e. Asymmetric recession of MR and LR with Y split of the Lateral rectus).

AA: Does not help a lot in globe retraction. But helps for the vertical shoots

SK: Agree. No sense to Y-split if retraction severe. You have to recess the lateral way back (supramaximal) or use periosteal fixation to reduce the retraction that will simultaneously relieve the up/downshoots.

DH: The recession alone helps reduce the upshoot. There is debate about how much the Y split adds once you have recessed the muscle. See <https://www.ncbi.nlm.nih.gov/pubmed/25727590>

SD: Yes, you can do simultaneously recession and Y splitting.

YM: Interesting idea. Haven't tried it

RK: Y split reduces the upshoot, but not retraction. If there is retraction we have 3 options; LR recession, differential recession of LR / MR or periosteal fixation, based on the severity of retraction.

10. In exotropic DRS, I have seen a few children develop upshoots, AHP and retraction as they grow up though primary gaze XT is small. What is the best approach in such cases? What age should we consider surgery?

VS: All these phenomenon (upshoots, globe retraction and AHP) are usually worse in patients with exotropic DRS. It is believed that it is the amount of innervation and tone of the LR muscle that decides the position of the eyeball in primary position as well as governs the associated phenomenon. Therefore, it is possibly important to look more carefully at these patients for all these phenomena, and the degree of co-contraction.

Possible options to manage these patients if the XT is small will be:

A) Only overshoots/ upshoots are significant components:

- Small LR recession with Y split
- Small LR recession with Y split/ IO surgery or SR surgery – if suspect associated innervational component

B) Upshoots and globe retraction all are significant:

- Asymmetric MR recession and LR recession in the same eye (MR: LR surgery 2:3 for orthotropia and Larger LR recession for exotropia in primary position) with Y split of LR

Age at surgery: When we can obtain reasonable measurements, as the primary position exotropia is small in question.

AA: Yes, I did. I would consider surgery whenever they develop a constant AHP or significant vertical shoots.

DH: The upshoot can be disfiguring and so when the patient becomes symptomatic (whatever the age), that is when I offer surgery.

SD: This is due to tight LR and I have done small LR recession with Y splitting in similar cases with good result.

RK: Best to perform surgery in the form of unilateral LR recession and Split (again depends on the other factors).

11. How do we predict co-contraction (except global retraction and shooting) in order to prevent consecutive XT?

VS: I think large amount of globe retraction and upshoots are the most important clues to detect the co-contraction.

AA: Perhaps doing a forced duction test on the lateral rectus muscle might give us an idea.

SK: Agree; but I think that assessing the retraction and up / downshooting *are important* in gauging the co-contraction, to prevent overcorrection in an eso DRS! In addition, I think that measuring the lid fissure height, as shown in the webinar, is a good gauge of co-contraction.

DH: Also watch for dysinnervation and avoid contralateral surgery in these cases; do not over-recess the medial rectus muscle. Intervene if this is evident in the early post-op period as it is not going to improve. Once an ET Duane goes XT it is very hard to get them back.

KA: I like to look at saccadic velocities, but centrifugal and centripetal, comparing the sound and involved eye. The adducting saccade of the involved eye is slowed and hypometric, with a noticeable post-saccadic drift. This occurs due to the co-contraction of the ipsilateral LR, and might be used to estimate the degree of co-contraction.

SD: Preoperatively, I look carefully of contralateral medial rectus adduction when effected eye attempting maximum abduction, if there is contralateral MR adduction deficit, albeit however small, I would be very cautious about dosage of bilateral MR recession, always aim to be a bit conservative to avoid over correction, especially in adult patients. Another tip is intraoperative FDT, if the ipsilateral lateral rectus is tight and MR not so much be careful of overcorrection.

YM: Do cover test for distance in adduction. If you get XT instead of ET – be careful. [J AAPOS. 2011 Aug;15\(4\):326-30. Doi: 10.1016/j.jaapos.2011.05.006.Risk factors for consecutive exotropia after vertical rectus transposition for esotropic Duane retraction syndrome. Velez FG¹, Laursen JK, Pineles SL.](https://doi.org/10.1016/j.jaapos.2011.05.006)

RK: Preop exotropia in adduction and near distance disparity are two important things. Be conservative with the amount of MR recession (no more than 5 mm) you do.

12. How useful would it be to perform a forced degeneration test by Romero Apis to know the amount of co-contraction? Will that help choose the surgical procedure accordingly?

VS: I don't have any personal experience with this test.

AA: Theoretically it might work, but never tried it before.

SK: I think viewer meant "force generation" test. It is useful in a cooperative patient. Have tried it in occasional cases in adults when I am not sure how active is the lateral rectus innervation on adduction. But with clues from answer to question 11, I can usually figure out how active is the lateral is and plan surgery accordingly, and rarely have I had to use that extra test.

KA: Same answer as for Q 11. Saccadic velocities might be easier and more comfortable for the patient. But forced generations should help estimate co-contraction.

YM: Sounds like a good idea.

RK: Interesting. But not sure how it can help in choosing a surgical procedure.

13. Respected Dr. Steve, do you intervene in ortho DRS to improve abduction?

VS: Specific question to Dr. Steve, but possibly these are not ideal patients for this indication.

SK: No. Any intervention to improve abduction risks compromising adduction, and that creates a new problem for the patient.

DH: I find that these patients have unrealistic expectations about what can be accomplished with surgery, no matter how much you tell them that it cannot be made normal. I have tried SRT in a patient who had a very small ET in primary gaze and while I was pleased that he did not have an adduction limitation or XT in primary with slight improvement in abduction, he was not satisfied.

RK: No. Best to avoid. SRT is a powerful procedure, which can lead cons XT.

14. Is SRT contraindicated in presence of severe upshoots?

VS: Possibly this question is specific to Dr. Hunter; however, I would avoid SRT in severe upshoots and severe globe retraction, significant limitation of adduction pre-operatively.

AA: For me yes, I won't do it in such cases

DH: It would need to be done in combination with surgery for upshoot.
See example case at <https://www.youtube.com/watch?V=d3brerlbzua>

SD: Not necessarily, if you do concurrent MR recession and SRT.

YM: I would be very careful and might recess the LR a bit before transposing the SR.

RK: Yes, in very severe cases. This is very rare. In over 90% of ET Duane cases, SRT is possible (mostly in addition to MR recession).

15. How does Dr. Hunter avoid ptosis in SRT?

VS: Specific question to Dr. Hunter, but I feel that unless we dissect the Superior rectus too far behind, we do not get ptosis in any SR surgery including SRT. It is more important to separate the Frenulum between SR and SO muscles, that can cause induced vertical squint / torsion.

AA: Ptosis is quite uncommon with meticulous dissection of the superior rectus muscle from the levator muscle.

DH: Agree. Ptosis has not been an issue.

YM: There is no ptosis after SRT

RK: Never seen any ptosis after SRT.

16. I definitely generate about 10% vertical strab when I do transposition surgery for sixth nerve palsy, which is why I have never adopted transposition for DRS treatment.

VS: For some reasons, this is more common with SRT for sixth nerve palsy, but we all don't see much induced vertical squint in DRS. There is not much induced vertical squint in primary position/ small vertical phoria which the patients tolerate well. They might have some vertical squint in attempted abduction but again it is not significant/ causes diplopia. It is worth trying.

SK: I have had 2 cases of induced verticals in DRS after VRT, among my first 4 cases. So I have not done any more – there are other options that work well for me with minimal risk of post-op verticals.

But Drs. Hunter (and his colleague Linda Dagi), Morad, and Kekunnaya have reported good experience with VRT in both conditions and they would be in much better position to answer the question.

DH: 6th nerve palsy is not the same as DS. Many of those patients have other nerve palsies so you have to be careful there. We have found 7% new verticals, but these can be managed and patients are so pleased with the improvement in abduction. You can be sure there will be no improvement in abduction with an MR recession, and because the MR recession is larger you have a greater risk of creating problems with adduction and convergence.

SD: I have done over 50 adults SRT for 6th nerve palsy and only one with postop vertical of 4PD which resolved by 6 weeks follow up. I found SRT is more effective in 6th palsy than in Duane ET patients. The key is to make sure the transposed SR maintains its normal anatomical distance from limbus & the tendon spread maintains its normal width. (personal experience due to my neurophthalmology bias as I don't think all the muscle fibres are innervated exactly the same for a given muscle for primary, secondary and tertiary functions of the muscle).

YM: For some reason SRT causes more vertical in 6th nerve than in Duane but it is very rare anyway

RK: I have not seen this with 6th nerve palsy. In our published series one patient developed small hypotropia; this can be managed.

17. Would a three muscle recession in first sitting have helped in this case; LR rec and MR rec of LE and MR rec in RE?

VS: It is not clear about which case. Possibly about the case with co-contraction case by Dr. Morad. Yes, possibly it is agreed that we could do asymmetric small MR recession in BE with LR recession, but it is in hindsight a learning lesson for managing patients with co-contraction.

AA: Yes, it might.

SK: If referring to the case of Dr. Morad, this 3-muscle approach is one I have done many times as a primary surgery to correct a combination of severe retraction and esotropia. For the eso, you recess the MR of DRS eye. To treat the severe retraction, you have to do very large recess of the LR in the same eye – which reduces the effect of the MR recess in treating the eso, so you have to add the MR recess in the fellow eye to correct the rest of the eso. With adjustable sutures this is a safe and effective option.

DH: I think unilateral MR recession and SRT would have been better!

SD: Possible.

RK: I would perform SRT and MR recession in these kind of cases.

18. How you calculate the amount of resection or recession in DRS, as they tend to overcorrect?

VS: In general, we all should target less amount of muscle recession for both MR and LR. If planning only MR recessions, do not exceed more than 5.5 to 6 mm and if doing MR recession along with SRT, possibly maximum of 4 / 4.5 mm MR recession is ideal.

For resection, Dr. Kraft already explained specific set of indications and amount of recession/ resection.

AA: I go for the standard tables, but avoid exceeding 5.5 mm medial rectus recession in primary cases, but might do more in reoperations

SK: Agree. I don't recess the medial more than 5.5 - 6.0.

And I emphasized that if the case fits the pattern where an LR resect is safe to do, then the amount of LR resect has to be small – around half of what one would normally do for a usual esotropia of that angle. That reduces the risk of post-op exotropia.

DH: Agree. For a standard SRT for an ET of about 30 in primary, I recess the MR 4.

YM: MR rec I do less than the tables about 0.5 - 1 mm. LR recession usually don't overcorrect in XT Duane.

RK: In most cases, 4 mm should suffice when you do along with SRT. Very rarely you might have to do 5-5.5 mm when the ET in PP is very large.

19. We reported using plication and partial tendon transposition in esotropic Duane. No vertical or torsional issues and blood flow maintained. Anyone else use that novel approach?

VS: It is an interesting idea and should work. But I have not tried. Will be worth trying.

AA: No, never tried it before.

SK: Never tried it either. But that seems to be a safe alternative – spares ciliary circulation.

DH: I avoid partial transpositions because they make too much scarring. Also you cannot be sure you have preserved blood vessels when you split the muscle.

See <https://www.ncbi.nlm.nih.gov/pubmed/30586616> for a discussion of this topic.

RK: No, I haven't done that.

20. I have a question for Dr Hunter: Do you think after SR transposition, the abduction improves over time? I have two cases that the abduction was -4 before transposition and after 5 years of PO, they have -1 and I see it improving year after year.

VS: Although specific question to Dr Hunter, I agree that it improves but most of the improvement happens until about 6 to 9 months. Usually not further change takes place beyond this time (and not beyond 12 months). This is common to all transposition procedures.

AA: Yes, abduction tends to improve over time but not in all cases.

DH: Abductions seems to improve slightly over time, though I wonder whether adduction limitation also increases over time.

We reviewed our data on this in <https://www.ncbi.nlm.nih.gov/pubmed/24763870>

Here is one example (all images show left gaze only, pre-op, 6 months' post-op, and 34 months' post-op):



YM: Could it be due to stretched scar of the MR?

RK: Yes, abduction improves over time.

21. In DRS with absent abduction, how much of hyperopia would be worth a prescription?

VS: I think any hypermetropia above 2.5 D for sure should be fully corrected in esotropic DRS, and will try to re-assess with glasses for HM more than 1.5 D.

AA: 3 or more hyperopia.

SK: Depends if there is also an esotropia in primary position!

A) If not, then I do not give Rx unless over +4 D, to prevent ametropic amblyopia.

B) If there is an esotropia in primary position then I treat as for any possibly accommodative esotropia: +2.00 D or over.

DH: I will sometimes give even +1.50 D depending on the family preference, likelihood to wear glasses, degree of head turn. But at +3 or above it is definitely worth trying. I've had patients with even more hyperopia come in for a second opinion after being offered surgery and they did quite well with glasses. At the very least it buys time.

KA: The answer will also depend on the primary position measurement. In those patients I've seen with an accommodative component, the angle in primary has been moderate to large. For an ET ≥ 15 with hyperopia of at least +2.50, it could be worth trying glasses. But in my experience, the cases with an accommodative component typically have +3.00 or more.

SD: Depends on the age of the child and I would trial anything over +2.0 dioptres

YM: +2.5 or more. Worth trying

RK: +1.50 D or more is worth trying in infants and toddlers with ET Duane. Please see: [Kekunnaya R, Velez FG, Pineles SL. Outcomes in patients with esotropic duane retraction syndrome and a partially accommodative component. Indian J Ophthalmol.2013 Dec;61\(12\):701-4.](#)

22. Any options for ortho DRS in order to improve abduction? My patient is a 40-year-old dancer, and is very keen to improve abduction.

VS: I think it is riskier to try to do surgery in such patients for this objective. We might be doing more harm by being more aggressive to such a patient. Rather important indications will be significant upshoots, and significant globe retraction.

AA: I would be afraid that I might induce diplopia and I would refrain from surgery.

SK Agree. See question 13.

DH: If this is the person's profession or avocation then after a very careful review of risks and benefits I would consider offering a procedure, explaining that we might have to go right back to the operating room and reverse everything, and see also comment above about these patients expecting more than can be achieved.

SD: You can consider SRT alone without doing MR weakening and it is generally safe though need consent the possibility of overcorrection. You can do adjustable suture SRT transposition and it is easily undone if any problem.

YM: I would hesitate to do surgery on an adult like your patient. The risk of post op diplopia is big

RK: I wouldn't risk any surgery on her.

23. Does Kyle use prisms to straighten the head?

DH: I think Kyle answered this question, but consider that even a very heavy, unsightly and expensive 12 PD prism is only going to improve head position by 6 degrees. A more realistic amount of prism, say 5 PD, only reduced the head turn by 2.5 degrees.

KA: Straightening the head can be achieved with prism glasses in some cases, but the number of patients to which this would apply (as an alternative to surgery) is small. The conditions have to be just right! This would work best in patients who are capable of bi-fixation and have smaller angles in primary. Large amounts of prism over one eye tend to be uncomfortable (and costly), and the alignment results of splitting the prism between the two eyes can be unpredictable due to variations in innervation patterns of the affected eye. Prism over the unaffected, fixating eye will move that eye out of primary position, with resulting changes in innervation to the Duane eye based on Hering's Law. Splitting the prism may be effective, but there is no algorithm with predictable results because each Duane case is unique. It is done by trial and error and prism power may need to be split asymmetrically.

24. What is the new classification for Duane syndrome?

VS: The clinical classification should include both Huber's classification as types I, II and III and subtype based on deviation (esotropia, exotropia and orthotropia). Type IV is synergistic divergence. Obviously all other factors such as globe retraction, AHP, degree of overshoots help in planning intervention.

AA: There are several ways to classify Duane syndrome, but for clinical purposes, they can be classified according to the alignment in the primary position, rather than the direction of limitation.

SK: Agree. I recommended no classification:

I made the point at start of my talk to not classify DRS. Just as was discussed in webinar 1 about congenital fibrosis of the EOM—Dr. Sener advised not to classify CFEOM either.

Both are congenital cranial dysinnervation disorders (CCDD) with a wide spectrum of clinical features. Use the 7 criteria in my talk to decide on treatment plan for DRS. It is more important to carefully examine the patient and decide on the problems that need addressing and their severity, rather than trying to assign a patient to some classification scheme.

DH: Agree. I think Steve's structure looking at the combination of primary position gaze, adduction limitation, abduction limitation, and dysinnervation is very helpful. What you end up with is about 300 types of DS instead of 3!

KA: Duane syndrome should be classified based on both, deviation in primary AND direction of greatest limitation on ductions. Type I Duane syndrome should be subdivided into ET Duane I, XT Duane I, and ortho Duane I. Same could

be said for type III: 3 subtypes based on primary position deviation. Type II, in my experience, tends to be universally exotropic in primary.

SD: Duane syndrome is considered a type of CCDD

YM: The old classification is worthless as this is a spectrum. In each patient there is a different blend of the following three elements: reduction in abduction of the LR, reduction of adduction of the MR and co-contraction of the LR.

RK: Classification doesn't help in surgery; Primary position and side gaze deviation, amount of AHP, grading of retraction, overshoot, amount motility limitation, field of BSV and patient's expectation decides the surgery. Some can be improved up to 100% and some only by 40-60%. So the criteria for successful strabismus surgery in Duane are different compared to typical comitant strabismus surgery.

25. Could Kyle discuss the role that orthoptists play in non-surgical management of patients with Duane's? What works? What does not work? Non-surgical ways to expand the binocular field? Etc.

KA: This would not be a short discussion! In my opinion, Duane syndrome is a condition best co-managed by the surgeon and the orthoptist. But if surgery is not an option for some reason, I would approach management this way. The first and most important step for the orthoptist is to manage amblyopia if present. Some of these cases have strabismic amblyopia, but others with strong co-contraction have anisometropic (meridional) amblyopia induced by the forces on the globe. Next step would be to restore a normal head posture using prism glasses or correction of moderate to high hyperopia. Exercises have limited efficacy because they primarily train the brain, not the EOMS. No amount of exercise can overcome the anomalous innervation and mechanical restrictions associated with the condition. The best potential (non-surgical) outcome would be bifoveal fixation, minimal to no head posture, and a small range of single vision around primary.

26. Is Duane Syndrome included in CCDD?

VS: Yes, it is well recognized and possibly an important part of CCDD.

AA: Yes

SK: Agree. See answer to 24 above.

DH: Yes, see our chapters on this in genereviews:

<https://www.ncbi.nlm.nih.gov/books/NBK1190/>

<https://www.ncbi.nlm.nih.gov/books/NBK1348/>

KA: Yes

SD: Yes

YM: Yes. See Parsa's paper quoted earlier.

27. What is the role of vertical muscle transposition to get abduction?

VS: Definitely VRT plays a major role in improving abduction in patients with esotropic DRS. Landmark studies by Dr. Rosenbaum's group and our experience show the greater improvement in abduction, and field of binocular vision.

AA: It improves abduction only slightly usually by 1 unit.

DH: In our original study we found improvement in abduction limitation from -4 pre-op to -2 post-op. In my experience families are almost always delighted to see this improvement. However, before surgery I say "we hope to get a slight improvement in the outward movement of the eye, which we know we will not see with the other approach (MR recession).

SD: In my experience it improves abduction deficit by 1-2 on the scale of 4 unless you do concurrent MR recession.

YM: Important.

RK: Yes, improves by 2units. Please see: [Tibrewal S, Sachdeva V, Ali MH, Kekunnaya R. Comparison of augmented superior rectus transposition with medial rectus recession for surgical management of esotropic Duane retraction syndrome. J AAPOS. 2015 Jun;19\(3\):199-205.](#)

28. What about results of SR transposition to improve abduction in Duane type 2?

VS: In my thought process, there is limited role for SRT in type 2 and type 3 DRS. Majority of patients with type 2 DRS (and even type 3 DRS) are exotropic. In exotropic patients, SRT has no role except when we combine LR periosteal fixation. However, even if there is esotropia with type 2 DRS, MR recession might be better. In select cases with no major co-contraction, upshoots and MR not tight, SRT should be tried.

AA: You mean adduction? If so it might increase the narrowing of fissure and the retraction. I never tried it.

DH: I have only used nasal transposition of the SR on one occasion and I had to take it down 3 days later because the patient complained of severe intorsion. However, Eric and Earl Crouch had an ARVO abstract on this where they claimed success in 2 patients: <https://iovs.arvojournals.org/article.aspx?ArticleId=2371654>

SD: You mean Adduction? Not as effective (one case only!)

YM: The problem in exotropic Duane is not the abduction usually.

RK: I haven't done that.

29. How do we diagnose Vertical Duane? Could you also elaborate on treatment modalities?

VS: Vertical Duane presents more like limitation of elevation and depression with variable degrees of retraction and palpebral fissure narrowing. Many of them have associated horizontal deviations. I have limited experience and feel that the goal should be to align in primary gaze and take care of associated overshoots/ retraction.

AA: Vertical Duane is when there is widening of the fissure on down gaze and retraction and narrowing of the fissure on upgaze with limitation of vertical movement. It is quite rare and requires vertical muscle recession.

DH: I think anything that you might want to call "vertical Duane" is just another CCDD and that you might confuse yourself to use that terminology.

RK: Lid fissure changes are seen on down and upgaze. There could be abnormal tightness of lid muscles as well. This is very rare form of Duane. Surgery depends the type deviation.

30. Dr. Hunter: When you adjust MRM to put back that means that do you left the eye with only SR transposition?

DH: That is correct. I will pull the MR all the way back up to the insertion. There are some cases where there is, say, just a very slight overcorrection, and if that occurs I might pull the MR from, say, 4.5 mm up to 2 mm.

31. Have you had any experience with Nishida procedure for DRS?

VS: Never tried, but worth trying it.

AA: I have used it for diseases other than DRS and find it very useful.

SK: Good question – I have not done it. That is why I asked Dr. Hunter – but he answered that he has not done it either.

DH: Not yet.

SD: I haven't done Nishida for Duane syndrome, but have done for double elevator palsy, MR transaction; they work very well.

YM: Not yet, but it sounds interesting

RK: No, but should work similar to SRT. We have done that in MED cases (instead of mono rectus transposition with good success).

32. Is the Forced degeneration test of Romero Apis carried out on the table, important to look for co-contraction?

VS: I do not have experience with this test.

AA: Yes, it might help.

SK: Agree. See answer to question 12.

DH: Have not tried it, but I just read about it in [Clinical Strabismus Management](#).

YM: For those who are not familiar with this test:



Figure 24-4. The force-degeneration test is an innovative new forceps force test suggested by Romero-Apis,¹⁴⁴⁻¹⁴⁹ to demonstrate lateral rectus (LR) anomalous innervation in opposite gaze in selected cases in which differential diagnosis needs clarification. Romero-Apis suggests the following, "To demonstrate co-contraction just with the forced duction test in those cases of substitution (no VIth nerve in lateral rectus muscle): (A) We ask the patient to look halfway between the primary position and adduction. Then, we try to move the eye into more adduction, and we feel positiveness because some restriction is originated by the lateral rectus co-contraction. (B) Again, we ask the patient to look halfway between the primary position and adduction, then we hold the eye with forceps, but now, we ask the patient to look outwards, and in the meanwhile, we move the eye inwards, and we feel negativeness (release of force), because the lateral rectus has been inhibited by its IIIrd nerve abnormal innervation." In other words, the forceps hold the eye in an opposite gaze position, not so far as to feel restriction but leaving some room for further forceps adduction. The patient is then instructed to change gaze toward the involved side (attempted abduction). The examiner, at this moment, feels a release of the anomalous LR co-contraction and can now rotate the eye still further inward. I would suggest that one might term this test the Romero-Apis force-degeneration test. Before this force-degeneration test is done, one has determined that there is no LR normal or (sub)normal active abduction innervation. The test determines the presence of anomalous LR innervation in opposite gaze.

It is done not on the table but when the patient is awake. Restricted LR on the table may imply severe co-contraction but also can imply fibrotic and weak LR.

33. I have found that majority of patients who undergo Duane syndrome surgery to correct an anomalous head posture are very happy with a reduction of their head turn to less than 10 - 15 degrees. Do you find that the increased abduction with SRT improves their quality of life enough to risk an overcorrection or inducing a vertical deviation?

VS: As discussed above, patients with esotropic DRS usually do not have much induced vertical squint and over-corrections if we limit MR recession (4 / 4.5 mm) and SRT. They usually have advantages of improved primary position deviation, abduction, as well as AHP. So, I feel that it is worthwhile to try in these type I ET DRS.

AA: I don't consider it my procedure of choice.

SK: Great point by this reader! Same answer for any strabismus surgery plan and result.

Surgery does not necessarily have to get the patient perfectly straight in primary position nor fully reposition the head to primary position. A small residual angle will still be a major benefit for the patient, and mild residual posture, under 10 degrees, should prevent osteoarthritis in the long-term. So agree completely that one should do an effective operation that addresses these issues, but not be so aggressive with the plan that you risk creating an overcorrection or new heterotropia.

DH: I have seen this infrequently enough that it has not been a major concern when endeavouring to do this surgery. I did not have a chance to discuss at the conference but I also put a short tag noose suture on the SR muscle and this gives another opportunity to adjust in the event there is a vertical deviation. One of the main reasons I have seen new verticals come in from elsewhere is that the SR is inadvertently recessed at the time it is transposed, or it is not attached along the Spiral of Tillaux. This will cause limited elevation on the side of the transposition.

SD: Agree with the comments on AHP correction and expansion of field of BSV. Vertical is rare.

YM: Yes, because it is improving the field of binocular single vision.

RK: Patients are really happy to see the improved abduction which is not seen with just MR recession. I used to do only MR recession for many years, but adding SRT provides 2 unit of improvement in abduction and improvement in the field of BSV. Why not give this additional benefit to the patients? I guess the risk of vertical deviation may be similar to the consecutive XT after larger MR recession.

34. What are the contraindications or better to avoid situations for performing SRT in ET Duane?

VS: I will avoid SRT in severe Upshoots and severe globe retraction, significant limitation of adduction pre-operatively.

AA: For me, a severe co-contraction evidenced by severe retraction on adduction and vertical shoots.

DH: If you are just adding this procedure to your surgical options then I agree you should avoid patients with patterns, upshoots, and pre-existing vertical deviations. However, over time each of these conditions can be managed. The main thing to watch out for is an adult who is going to have a negative reaction to XT in adduction.

SD: Avoid overcorrection by assessing contralateral adduction; use adjustable suture if you can.

YM: Severe retraction.

RK: Only in cases of ET Duane with very severe retraction / overshoot.

35. Can we perform only MR resection in small angle ET Duane?

VS: Do you mean only MR Recession? MR recession alone can be done and will be preferred if the MR is tight on intra-operative FDT. However, if the MR is not tight, only SRT can also be done.

AA: If you mean MR recession, then yes.

SK: Agree. If you meant recession – then if small angle (under 20 PD) of eso in primary position with a compensatory head turn posture, then single MR recess is still a good option to address the head turn and esotropia in primary position. However, as noted in the webinar, it does not lead to much improvement in abduction in many cases, if that extra benefit is a desired goal.

DH: Do you mean LR resection only? Potentially, but the MR is likely tight. I published one case where we did a 2 mm recession and 2 mm resection for a small-angle DS with atypically good abduction. <https://www.ncbi.nlm.nih.gov/pubmed/27965142>

SD: Presume you mean MR recession, yes, if it is what you meant.

YM: Since the MR is fibrotic I don't think this will work.

RK: If the viewer meant LR resection: I would do only MR recession as most ET Duane patients have tight MR which need to be tackled first.

36. When should we consider doing only MR recession in Duane?

VS: Small angle Esotropic DRS (< 20 PD) with tight medial rectus on FDT.

AA: This is the standard surgery in Duane, so practically in all cases of Duane type I.

SK: Agree. See answer to question 35 above. If small angle (for me 18 PD or less) it is still a good first step. It does not have high risk of creating new problems.

DH: I rarely perform this procedure in unilateral DS. However, in bilateral DS I think it is probably fine.

YM: Small ET with -4 abd. def.

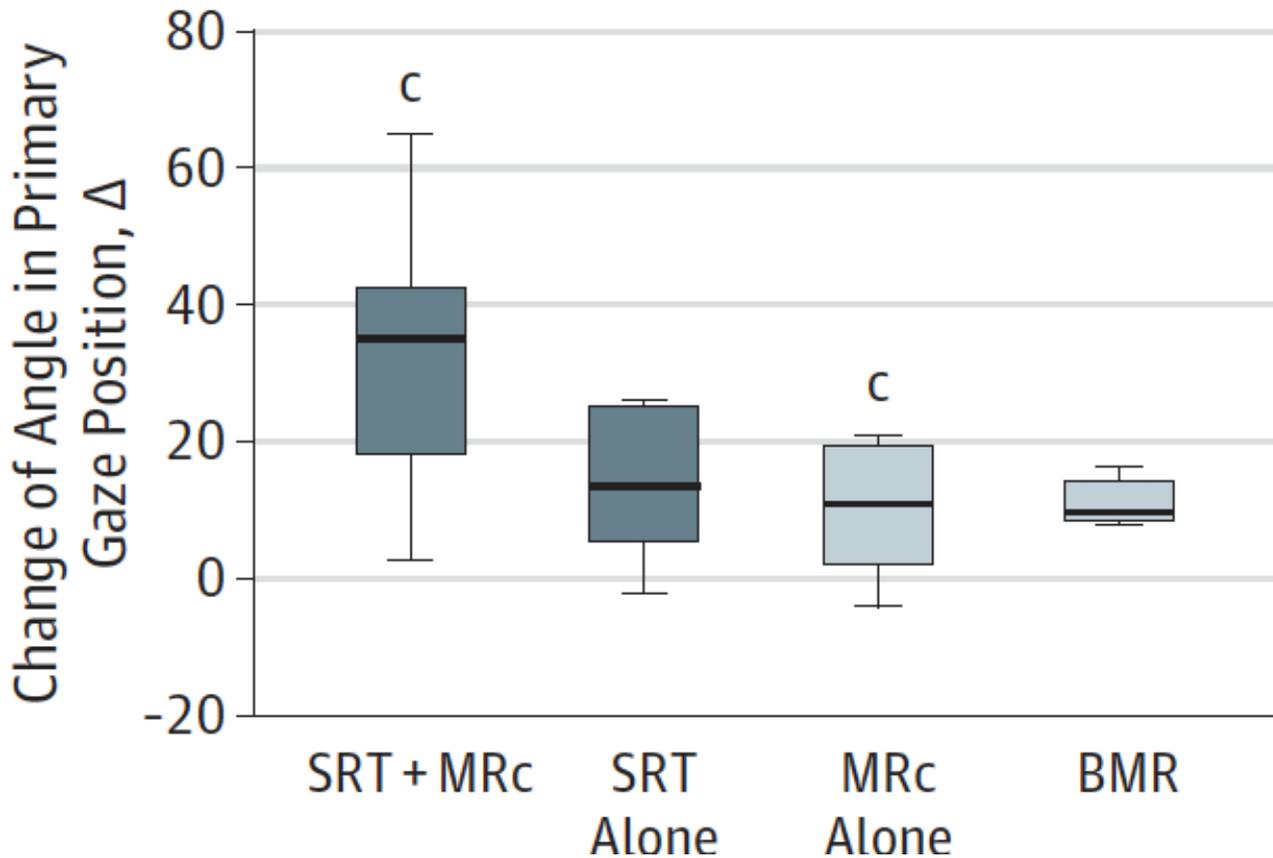
RK: Very tight MR with small angle and in the presence of significant retraction. If the MR is very tight, recessing itself can improve the abduction a bit (not otherwise)

37. How much correction does SRT provide?

VS: SRT with augmentation without MR recession corrects about 15 PD of esotropia. If done along with mrc of about 4 / 4.5 mm, we can get correction of about 35 PD of esotropia.

AA: Around 15 PD

DH: Agreed, we found 15 PD mean effect for SRT alone in our JAMA Ophthalmology article vs. 30 PD for SRT+mrc:



SD: For Duane syndrome, SRT alone corrects around 10PD but it can be up to 20-30PD if you add MR recession of various amount. In 6th nerve palsy SRT alone can correct 20PD in my hand.

YM: Around 15D

RK: Around 12-15 PD according to our study and also other published papers.

38. Any complications of SRT?

VS: Yes, small risk of induced vertical squint, over-corrections, and torsion. Can lead to significant large hypertropia if adequate separation from SO is not done.

AA: Vertical deviations and overcorrection.

DH: Agreed. One has to be alert for torsional diplopia but so far it has not been an issue.

SD: Rare, vertical deviation, torsional diplopia

YM: Over-correction with abd. def., vertical deviation (rare).

RK: Rarely vertical tropia and torsional diplopia.

39. Should SRT always be combined with Mrec?

VS: Most of the time patients with type I esotropic DRS have a tight MR. So, it is worthwhile to add MR recession. However, in patients when MR is not tight, or max 1 + tight on FDT, it can be performed alone for small ET of < 20 PD.

AA: Yes, in most of the cases, unless the medial rectus was not tight at all.

DH: See response above. You can correct about 15 PD of ET in primary with SRT alone.

SD: For most cases!

YM: Usually because the fibrotic MR will generally prevent abd.

RK: Yes, in most cases.

40. Any role of VRT now?

VS: Can still be done for Esotropic DRS especially large angle ET (say 45- 50 PD), though less common.

AA: Yes, in severe limitation with large angles.

DH: We have occasionally gone back after SRT to obtain additional effect with IRT.

YM: Not for me.

RK: Not done any in Duane, but have done it in sixth nerve palsies.

41. What is the recommended the distance between the 2 arms in Y split?

VS: The recommended distance between the 2 arms is at least the width of the LR, i.e. about 10 mm. However, in our experience, we increased the distance for larger grades of upshoots i.e. about 12 mm for grade 2+ to 3+ and about 14 mm for grade 4 overshoots.

AA: At least 10 mm (full muscle width).

SK: Agree. I place the segments (halves) just above and below the levels of the upper and lower poles, respectively, of the original LR insertion.

SD: I usually use the full tendon width of LR as a guide.

YM: 8 mm.

RK: 10 - 12 mm.

42. When do you do bilateral MR recession?

VS: We can do in 2 situations:

1. Bilateral esotropic DRS
2. Unilateral esotropic DRS with deviation > 20 PD. Can do bilateral MR recession with slightly greater amount in the normal eye if you don't want to do SRT

AA: For large angles over 25 PD and for severe co-contraction when you need to add lateral rectus recession and with very tight muscles creating fixation duress.

SK: Agree. It is one option when angle of eso too large for one muscle, and you do not want to do VRT.

DH: I would not perform bilateral MR recession for unilateral DS.

YM: For large angle ET Duane with severe retraction.

RK: Very large angle ET with severe retraction and tight MR.

43. Is augmentation a must for SRT?

VS: Not necessary in all patients but it increases the amount of correction in primary position deviation. So, if we have ET DRS of about 12 to 14 PD, we can do SRT alone, or in patient with ET DRS of about 30 PD, we can do MR recession with SRT alone (without augmentation).

AA: Not a must.

DH: No, in fact I probably only use it 30% of the time now, which may reduce the likelihood of new vertical deviations and makes it easier for the SRT itself to be adjustable.

SD: I add augmentation suture if abduction deficit >4.

YM: No, but it can help in improving the Abd.

RK: I always use augmentation suture. I think this is necessary to transfer the vector force from SR to LR.

44. Why not advance MR in consecutive XT?

VS: I think this is about the case of co-contraction. This was a specific scenario which was highlighted where co-contraction was significant. However, without significant co-contraction we can follow the following approach: MR advancement if there is significant limitation of adduction and intra-op FDT showing lax MR. However, if MR is not lax then the SRT might need to be tackled.

AA: Because the lateral rectus is very tight, so advancing the medial rectus might increase the co-contraction.

SK: I think that is an option, and I have had to do that in some cases. If LR is tight, as AA indicates, then I find you have to recess it as well to reduce the co-contraction that he correctly warns about.

DH: In the case presented with XT after BMR I would have advanced the contralateral MR.

SD: It is the tight LR that drives the over correction especially in case you done BMR recession.

YM: We can do that but sometimes only LR Rec will help.

RK: It is definitely an option, unless there is severe co-contraction and tight LR.

45. What are the specific indications for periosteal fixation in XT DRS? Is it reversible?

VS: Type II / Type III DRS with moderate or large angle exotropia in primary position, especially when associated with large overshoots / and globe retraction.

AA: Large angle with very tight LR and marked limitation of adduction. The classic indication for me is synergistic divergence. And yes, it is reversible.

RK: Very large XT Duane with very severe overshoot and Synergistic divergence.

46. Anyone faced consecutive ET in XT Duane?

VS: Touch wood not but residual XT many times.

AA: Quite rare. Undercorrection is more common.

SK: Agree.

DH: I have had patients come see me who were unhappy with the new ET in side gaze where they used to be XT in all gaze directions. So even if they are undercorrected in primary they might still be unhappy with ET in the field of action of the LR. Some have come back to then do SRT in those cases though I have not tried it yet.

SD: Not yet.

YM: Sure but usually it improves over time.

RK: No, I usually see undercorrection.

47. What are the options for residual XT in XT Duane?

VS: It depends on the following: amount of residual XT, primary surgery done and associated overshoots.

If small residual deviation/ no major Upshoots/ retraction: can do re-recession with Y split if not done before/ surgery on contralateral LR muscle

If large residual deviation/ no major Upshoots/ retraction: surgery on contralateral LR muscle

If large residual deviation/ and large Upshoots/ retraction: can consider periosteal fixation

AA: LR-rerecession or medial rectus plication, or LR periosteal fixation.

DH: Could also consider recession of the contralateral LR.

SD: Large contralateral LR recession often helps.

YM: Depends on what you did in the first surgery. More LR REC or more Mr res.

RK: LR re recession +/- MR plication.

48. How do you decide between srec and IO surgery in Hypertropia and Type 2 / 3 Duane syndrome?

VS: These are the possible general guidelines:

- Useful in patients with innervational upshoots, and associated moderate/ large vertical deviation in primary position > 10 PD
- Signs of Inferior oblique over-action, excyclotortion which suggest that IO weakening might be useful.
- If these are not there, we can possibly consider SR surgery
- If you suspect a mixed mechanism overshoot, following is useful:
 - We should look for the tightness of LR, SR and IO by intra-operative FDT/ traction test, and anomalous position of the LR/ SR muscle. Look also for any accessory muscles.
 - We recommend that if LR is tight, it should be tackled first followed by repeat FDT for SR and Traction test which might help decide which muscle can be tackled.
 - If there is a doubt in such cases, we can always do a staged procedure.

AA: Fundus torsion mainly.

DH: Agree.

SK: Firstly - do not classify the DRS!

I look at:

A) The clinical pattern of the hyper – is it large in primary position and is there a gradual updrift from abduction to primary position and into adduction. That suggests an innervational basis to the Hyper – and may be SR or IO co-innervation.

B) Size of the hyper – if over 10-12 PD in primary and no extorsion of fundus then I would consider the SR as possibly responsible.

C) If extorsion is present – then I would consider the IO may be responsible.

Also please see my answer to question 1 above.

SD: Fundus torsion: if there is significant excyclotorsion then consider IO weakening! I tend to avoid SR recession in Duane's patient as the vertical upshot often in adduction!

YM: Excyclotorsion= IO. Anomalous SR = SR rec.

49. What is the role of Prisms in Duane syndrome?

VS: I think specific question for Kyle. However, in general there is a limited role for prism in small angle ET / XT DRS without much upshoots, retraction and AHP but patient is symptomatic.

Similarly, in small angle consecutive ET/XT DRS after primary surgery and diplopia in primary gaze.

AA: Little role in my hands.

SK: I use them as a temporary measure – for patients awaiting surgery to control torticollis and / or diplopia. I use either ground-in prism if not large angle (under 10 PD), or Fresnel prism if larger angles. I also use ground-in prism long-term in patients with small angles who want to improve the head posture and reduce the deviation in primary position and also wish to avoid surgery.

DH: See comment above. 5 PD of prism over one eye corrects only 2.5 degrees of head turn. Anything more than that and you have a very heavy, expensive, and optically unacceptable prism.

KA: The answer depends on the patient's treatment goals. But generally, there are two situations where I would try prism. The first is a case with small primary position angle, in a patient capable of bi-fixation, who declines surgery. The second is the case of an over-correction. The latter is likely to need re-operation, but prism may make the patient comfortable in the short term.

YM: Only to improve diplopia post op.

RK: Only in cases of post op diplopia.

50. Is MRI a must in DRS?

VS: I personally feel that MRI is not a must in all cases of DRS. We might need to consider MRI for research purposes or for decision making in patients with significant overshoots. In such a case imaging should focus on orbital imaging to look for the Muscle position / bands.

AA: Not a must.

SK: Agree that it's not a must in most cases, but I would order it in one of two situations:

A) If the time of the onset of the ocular motility picture is not clear – and may be acquired. Most “benign” DRS cases will have a history of onset in childhood, usually early years.

I had one case many years ago of a “classic” unilateral eso DRS picture in an adult – and whom I had examined several years earlier and documented a normal motility exam! I ordered a CT of her orbits and it showed a meningioma that had invaded the posterior orbit. So aberrant innervation of the third nerve due to a slowly evolving lesion can cause a DRS-like picture.

B) For research or study purposes – to see either the static muscle structures (see work by Demer et al), or to confirm the sixth nerve agenesis or hypoplasia (see work by our South Korean colleagues), or else using a cine-MRI to show the dynamic changes in muscle calibre to confirm the co-innervation of one or more muscles with the lateral rectus (see work by Demer and originally reported by Cadera et. Al).

DH: I do not routinely obtain MRI, but I have seen cases of 6th nerve palsy misdiagnosed as DS including a young woman with abduction limitation and no globe retraction who had a brain tumor. So you want to be sure there is globe retraction before you call it DS.

KA: We know that some cases of Duane syndrome have ipsilateral heterotopic LR, and a high-resolution orbital MRI would identify these cases pre-operatively. But these cases also tend to have unusual alphabetical patterns such as Y or V. So there may be a clue to the placement of the LR found in measurements in secondary and tertiary positions of gaze pre-operatively. However, unusual patterns may also be caused by anomalous innervation of the LR by the vertical recti.

SD: Only if the pattern doesn't fit typical Duane syndrome as Duane syndrome can be mixed with other CCDD.

YM: No.

RK: Not a must, unless there is a diagnostic dilemma.