Transposition procedures in Duane retraction syndrome

Jefferson J. Doyle, MBBChir, PhD, MHS,[a,b,c,d] and David G. Hunter, MD, PhD[a,b]

SUMMARY Duane retraction syndrome, or Duane syndrome (DS), is one of several congenital cranial dysinnervation disorders. Patients present with limited horizontal eye movement(s) and globe retraction with eyelid fissure narrowing on attempted adduction due to co-contraction of the lateral and medial rectus muscles in one or both eyes. Various surgical approaches have been proposed to improve binocular alignment, reduce head turn, and minimize undesirable up- or downshoots in DS. Transposition procedures are one such approach, and a number of techniques have been described. These may involve one or both vertical rectus muscles and may or may not include full or partial disinsertion of the rectus muscle(s) from the insertion. Options involving both vertical rectus muscles include full vertical rectus transposition (VRT), partial VRT, rectus muscle union, and other modifications to be discussed. Options involving one vertical rectus muscle include superior rectus transposition (SRT) and inferior rectus transposition (IRT). The effectiveness of any transposition procedure may be enhanced with augmentation (posterior fixation) sutures, resection of the transposed muscle(s), and/or simultaneous weakening of the ipsilateral medial rectus muscle. This review discusses the indications, strengths, weaknesses, and other considerations of these approaches within the context of DS. Since the majority of DS cases are unilateral and most have the esotropic form, this will be the main focus of the review, although other forms will also be discussed. (J AAPOS 2019;23:5-14)

Duane retraction syndrome, or Duane syndrome (DS), is a congenital cranial dysinnervation disorder whose underlying pathophysiology is failure of the abducens nerve to fully innervate the lateral rectus muscle, with additional aberrant innervation from branches of the oculomotor nerve. This manifests as limited horizontal eye movement(s) in one or both eyes, with globe retraction with eyelid fissure narrowing on attempted adduction caused by co-contraction of the lateral and medial rectus muscles. Although Huber[1] classified DS into three types, for surgical planning it is often more straightforward to categorize it as orthotropic, esotropic, or exotropic. Various approaches have been proposed to improve binocular alignment, reduce head turn, and minimize undesirable up- or downshoots in DS. Transposition procedures are one such approach, and a number of techniques have been described. This review will discuss the indications for (as well as the strengths and weaknesses of) these approaches within the context of DS. Because 85%-90% of DS cases are unilateral and most have the esotropic form, this will be our main focus.

Unilateral Esotropic DS

The indications for surgical intervention in esotropic DS include the presence of a compensatory head posture toward the affected side that is large enough to cause symptoms (neck discomfort, need for preferential seating, or poor detection of objects in the ipsilateral visual field), esotropia in primary gaze that interferes with normal eye contact, or loss of control of alignment sufficient to cause reduction of binocular vision or amblyopia. Horizontal surgery for DS is relatively simple, usually fairly predictable, and conveys minimal risk for anterior segment ischemia (ASI).[2] However, this approach does not improve abduction, invariably limits adduction, does not improve (and may reduce) the range of binocular single vision (BSV), and may have a shorter duration of benefit.[3] For this reason, many surgeons advocate for the use of transposition procedures almost exclusively, unless the angle of deviation in primary gaze is small ($<10^{\Delta}$), preoperative abduction is −2 or better, and globe retraction is minimal, in which case a recession with or without judicious

Author affiliations: [a]*Department of Ophthalmology, Boston Children's Hospital, Boston, Massachusetts;* [b]*Department of Ophthalmology, Harvard Medical School, Boston, Massachusetts;* [c]*Division of Pediatric Ophthalmology, Wilmer Eye Institute, Johns Hopkins Hospital, Baltimore, Maryland;* [d]*Institute of Genetic Medicine, Johns Hopkins University School of Medicine, Baltimore, Maryland*

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Correspondence: David G. Hunter, MD, PhD, Boston Children's Hospital, 300 Longwood Avenue, Fegan 4, Boston, MA 02115 (email: david.hunter@childrens.harvard.edu).

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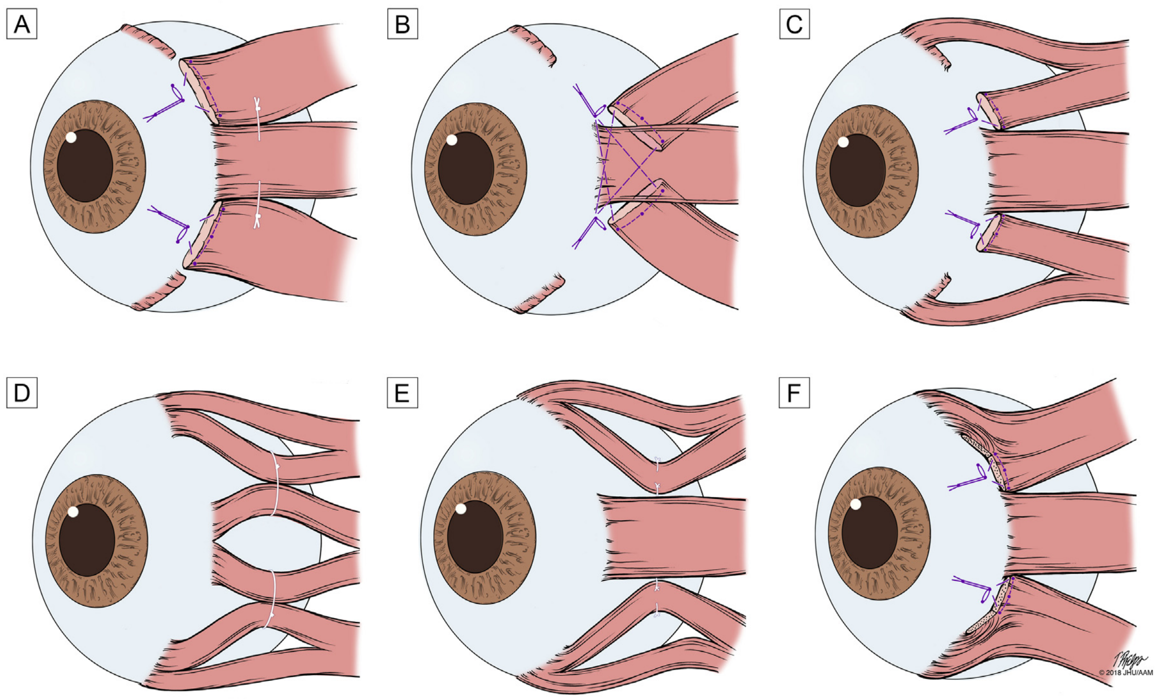

FIG 1. Variety of approaches to vertical rectus transposition (VRT). (These renditions depict optional short tag noose adjustable sutures and augmentation (posterior fixation) sutures where appropriate.) For partial and split tendon approaches, care is taken to avoid ciliary vessels when possible. A, Full VRT, adapted from Del Pilar and Kraft,[7] with lateral placement of the superior and inferior rectus muscles along the spiral of Tillaux. B, Crossed-adjustable technique, adapted from Phamonvaechavan and colleagues.[8] The transposed vertical rectus muscles are passed beneath the lateral rectus muscle and attached at the respective opposite corners of the lateral rectus muscle insertion. C, Partial VRT, adapted from Brooks and colleagues,[9] with placement of the lateral halves of the vertical rectus muscles adjacent to the lateral rectus muscle insertion along the spiral of Tillaux. D, Jensen rectus muscle union, adapted from Jensen.[10] The superior, lateral, and inferior rectus muscles are each split. Sutures are then used to tie the superior and inferior rectus muscles to the superior and inferior halves of the lateral rectus muscle belly, respectively, near to the globe equator. E, Partial VRT, without disinsertion of the vertical rectus muscle insertions, as proposed by Nishida and colleagues.[11] The vertical rectus muscle bellies are split longitudinally from the center of the muscle insertion for about 15 mm. The separated lateral bellies of each vertical rectus muscle are attached to sclera adjacent the respective border of the lateral rectus muscle 8 mm posterior to its insertion, using two sutures for added stability. F, "Full" VRT with ciliary-sparing partial resection of the vertical rectus muscles, as proposed by Hendler and colleauges.[12] An incision of approximately 2.5 mm is made perpendicular to the path of the superior rectus muscle on its temporal side at a predetermined distance (3-4 mm) the muscle insertion. An absorbable suture is woven through the posterior part of the transected portion of the muscle and used to transpose the entire muscle by reattaching it to sclera at the superior border of the lateral rectus muscle insertion along the spiral of Tillaux. This is then repeated for the inferior rectus muscle.

resection may be preferred. Although Jampolsky[4] states that transposition is "contraindicated if there is severely anomalous lateral rectus function," with use of adjustable sutures it is possible to manage an overcorrection secondary to residual or anomalous lateral rectus function accordingly.

Transposition procedures may involve one or both vertical rectus muscles and may or may not involve full or partial disinsertion of the rectus muscle(s) from the insertion. Options involving both vertical rectus muscles include full vertical rectus transposition (VRT), which involves displacement of both the superior and inferior rectus muscles to the insertion of the lateral rectus muscle,[5,6] partial VRT, rectus muscle union (Jensen procedure), and other modifications. Options involving one vertical rectus muscle include superior rectus transposition (SRT) and inferior rectus transposition (IRT). The effectiveness of any transposition procedure may be enhanced with augmentation (posterior fixation) sutures, resection of the transposed muscle(s), or simultaneous weakening of the ipsilateral medial rectus muscle.

Full Vertical Rectus Transposition (VRT)

Full VRT can improve the esotropia, head turn, and abduction deficit in esotropic DS (Figure 1A), even in patients who have failed prior nontransposition surgery.[13] VRT also improves the range of BSV on average about twice as much as isolated medial rectus recession.[13] A major factor affecting the efficacy of VRT in esotropic DS is the degree of tightness of the ipsilateral medial rectus muscle, which can contribute to postsurgical undercorrection or recurrence if not addressed.[13] This is even more important in DS than other causes of esotropia (eg, abducens nerve palsy), because the medial rectus muscle in DS can be fibrotic and stiff. During intraoperative forced ductions, the point in abduction where restriction begins approximately indicates the maximum amount of abduction to be gained from transposition alone. If more abduction is necessary, then combined medial rectus weakening should be strongly considered.

Full VRT Augmented by Ipsilateral Medial Rectus Weakening

A reduction in medial rectus muscle strength can be achieved by both surgical and pharmacological approaches. When disinserting and transposing both vertical rectus muscles in the case of full VRT, simultaneous recession of the ipsilateral medial rectus muscle is generally not performed because of concern for ASI. An alternative approach to weakening the muscle without compromising its blood supply is to use botulinum toxin, which may produce the same magnitude of effect as medial rectus recession but without increasing the risk of ASI.[14] It can be performed either at the time of surgery or 5-15 days later, with no difference in effectiveness, induced ptosis, or vertical tropia between the two time points.[14] If the patient can tolerate it, delayed injection in clinic at the postoperative visit may be preferable in adults, because it allows for the response to surgery to be assessed before applying treatment that might otherwise prove unnecessary or unhelpful. Another advantage of botulinum toxin injection is that it can be repeated postoperatively if the initial magnitude or duration of effect is inadequate. We are unaware of any reports of outcomes of botulinum-augmented VRT in the context of DS.

Full VRT Augmented by Ipsilateral Lateral Rectus Plication

If a patient has residual ipsilateral lateral rectus muscle activity, some advocate for a muscle-tightening procedure, such as a resection or plication. Resection is not advisable at the time of full VRT because of the risk of ASI; plication, however, can (but does not always) spare the anterior ciliary vessel(s).[15] Other reported advantages include the shorter operating time, early reversibility, and less surgical trauma, inflammation, and bleeding.[15-17] Lateral rectus plication has been shown to improve the esotropia and abduction deficit in esotropic DS in combination with VRT, with or without simultaneous medial rectus weakening if it is found to be tight on forced duction testing.[18] Although this approach may be a useful surgical option for esotropia from abducens nerve palsy, it should be used with caution in esotropic DS, because almost all patients show some adduction limitation on preoperative examination and lateral rectus muscle tightness intraoperatively that one would not wish to exacerbate.

Full VRT Augmented by Posterior Fixation Suture

A posterior fixation or augmentation suture ("Foster augmentation"),[19] is usually placed in the sclera 12-16 mm posterior to the limbus and adjacent to the lateral rectus muscle, incorporating one-quarter to one-third of the transposed vertical rectus muscle (Figure 1A). Augmentation sutures can enhance the effect of VRT in DS on the esotropia and head turn,[13] reduce the rate of additional surgery for undercorrection,[20] and decrease the risk of induced vertical deviation.[13] Augmentation suture placement may enhance the effect of a previously performed VRT when added later.[19] However, the data suggest that augmentation provides no added benefit unless the primary position deviation is $> 20^{\Delta}$ or the initial head turn is $> 20°$[13,20]; hence, augmentation sutures may best be reserved for cases in which either of these criteria are met. High-resolution magnetic resonance imaging studies support Foster's hypothesis that augmentation suture placement achieves its effect by displacing the vertical rectus muscle pulleys (and hence the muscle forces) into alignment with the new insertion.[21]

Full VRT Augmented by Transposed Muscle Resection

Rao has suggested that augmentation sutures may work by increasing the path length of the VRT muscles (Rao R. Variations in transposition surgery. AAPOS Annual Meeting, Charleston, SC, 1997). If true, a simple resection of the transposed muscles should achieve a similar outcome. Indeed, combining VRT with a 4-8 mm resection of the transposed muscles can enhance its effect, at least in paralytic strabismus.[7,9] Resection does offer some advantages over augmentation sutures, including avoidance of additional scleral passes, the opportunity to perform the transposition in a quantitatively graded manner (although validated dose–response curves do not yet exist), the ability to correct a preexisting vertical tropia by differential resection of the two vertical muscles, and compatibility with adjustable sutures.[12] Whether this approach proves to be a useful addition to our surgical armamentarium for DS remains to be determined.

Full VRT Augmented by a "Crossed-adjustable" Approach

Using a "crossed-adjustable" technique to augment VRT also increases the transposed muscle path length while avoiding resection of the transposed muscles (Figure 1B).[8] In the original description for abducens nerve palsy, it reduced the need for simultaneous ipsilateral medial rectus weakening more than twofold. Another advantage is the ability to employ an adjustable suture technique. The broad applicability of this technique in DS remains unexplored.

VRT Complications and Risk Factors

Complications associated with VRT include undercorrection, overcorrection, new-onset symptomatic induced vertical deviations, and ASI. Preoperative risk factors for undercorrection include greater esotropia in primary position (and in adduction) and greater tightness of the medial rectus muscle (with more restriction to abduction on forced duction testing).[22] Overcorrection may occur whether the transposition is performed in isolation or in combination with delayed ipsilateral medial rectus recession. Risk factors for overcorrection include younger age (about 2 years), smaller preoperative esotropia at near and in adduction, and less restriction on forced ductions.[22] Indeed, exotropia in adduction indicates that the ipsilateral lateral rectus muscle has significant co-contraction or tightness that will exaggerate the effect of the VRT in adduction. Another risk factor for overcorrection is simultaneous contralateral medial rectus recession, which can cause exotropia from prolonged fixation duress. Finally, a preoperative accommodative component to the esotropia can lead to the surgeon's overestimating how much surgery is needed to correct the esotropia. An accurate refraction should be performed and adequate optical correction provided before finalizing the surgical plan; indeed, for some patients full optical correction alone may be sufficient to adequately correct the esotropia and head turn.

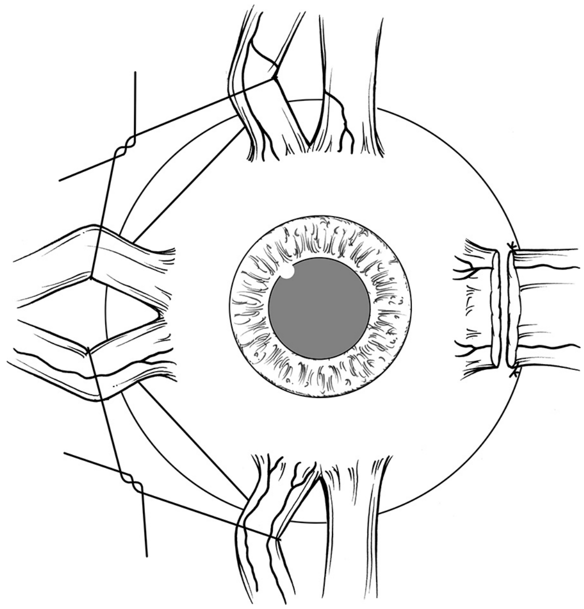

FIG 2. Variation in anterior ciliary vessel topography; adapted from von Noorden.[34] Only a single anterior ciliary vessel may be encountered in the distal aspect of the muscle that then branches into two or more arteries close to the muscle insertion (as illustrated here for the superior rectus muscle). Alternatively, one or two of the ciliary arteries may be located in the central aspect of the muscle, rather than the marginal aspect as is generally assumed (as illustrated here for the inferior rectus muscle). In either scenario there is a risk of incorporation of both anterior ciliary arteries of a given vertical rectus muscle being included in the transposition.

VRT Complications: Vertical Tropias

Hypotropia of the operative eye with restriction has been reported in 6%-30% of patients after VRT.[20,23,24] It can reportedly be minimized by meticulous surgical dissection of the attachments between the inferior rectus muscle, the capsulopalpebral fascia, and Lockwood's ligament. Induced vertical or cyclotropias from altered tension or direction of action of the superior oblique muscle can also occur; thus dissection of the attachments between the superior rectus muscle and the superior oblique tendon are also important.[25] Intraoperative forced ductions can help identify patients at risk for a new-onset hypotropia.[23,26] Restriction of elevation can be addressed by removing any augmentation suture that is attached to the affected muscle or recessing the tight muscle up to 3 mm from its transposed site. If still positive, other issues should be explored (eg, attachments to the superior oblique or inferior rectus muscles).

An early modification of VRT designed to reduce both induced vertical deviations and under- or overcorrection was the introduction of an adjustable suture technique,[6,27] which allows postoperative recession of the appropriate vertical rectus muscle(s) to correct any induced vertical deviation as well as reversal of horizontal overcorrection by recession of both transposed muscles from their new points of insertion. There are a number of adjustable suture approaches,[28] several of which have been used in both adults and children.[29] The short tag noose technique allows modulation of the outcome for up to a week after surgery, hence combating any immediate or short-term delayed horizontal or vertical misalignments.[30] In adults, delayed postoperative adjustments can be performed in the office; in children, they are usually performed in the operating room under brief, masked sedation. Although placement of an augmentation suture limits the ability to adjust the position of the muscle, it does not completely eliminate it.

VRT Complications: Anterior Segment Ischemia

ASI is a rare complication of VRT. Patient risk factors associated with ASI include advanced age, vascular disease, diabetes mellitus, hyperviscosity, hemoglobinopathies, and 360° scleral buckling.[31] Limbal incisions may also increase the risk.[27] The potential of vision loss from ASI has inspired development of a range of modifications to VRT as well as a number of other transposition procedures.

One method for reducing the risk of ASI with VRT is to transpose only the outer two-thirds of each vertical rectus muscle (so called partial VRT, or the Hummelscheim procedure; Figure 1C).[32] Although partial VRT may or may not be as powerful as full VRT,[13] it is designed to preserve one of the two ciliary arteries in each muscle for patients at risk of ASI, theoretically permitting additional surgical intervention on other rectus muscles, for example, recession of a tight ipsilateral medial rectus muscle. It is also compatible with resection, placement of an augmentation suture, and use of adjustable sutures. However, the low incidence and idiosyncratic nature of ASI has made it difficult to definitively demonstrate that partial VRT reduces the risk of ASI. Furthermore, some surgeons avoid using partial transpositions because increased scarring can result from longitudinally splitting a muscle, which can create adhesions to the globe along the full length of the split muscle.

Rectus muscle union (Jensen procedure), originally described in the context of abducens nerve palsy (Figure 1D), may also spare ciliary vessels.[10] With the lateral rectus muscle and both vertical rectus muscles split and sutured together without having been disinserted, the anterior ciliary arteries of each rectus muscle are theoretically preserved. Favorable outcomes have been described in abducens nerve palsy[33]; data in DS, have not, to our knowledge, been published. Of note, ASI has been described even with the Jensen procedure.[33,34] Possible explanations include inadvertent severing of anomalous anterior ciliary arteries during muscle splitting or severing/strangulation of the vessels during suture placement or tying. The topography of the anterior ciliary arteries can vary greatly, and careful identification of their location and course prior to muscle splitting may help reduce this risk (Figure 2).[34] Kushner[35] has proposed a modified Jensen procedure to preserve anterior segment blood supply by avoiding manipulation of the lateral rectus muscle vasculature.

One concern with the Jensen procedure is the potential redirection of some of the residual abducting force of the lateral rectus muscle during its splitting and attachment to the vertical rectus muscles, which may be counter to the surgical goal of increasing the abducting ability of the eye. An alternative approach is to split the vertical rectus muscles as one would for a partial VRT but to leave the muscles attached, then to place a suture 8-10 mm posterior to the insertions and transpose the muscle bellies by suturing them to sclera adjacent to the lateral rectus muscle (Figure 1E).[11] This helps preserve the ciliary circulation, avoids compromise of any residual lateral rectus muscle function, and permits simultaneous ipsilateral medial rectus recession. While outcomes for this technique have been described for abducens nerve palsy, there does not appear to be data for DS. As described, this approach precludes use of an adjustable suture technique, although simultaneous medial rectus recession with placement on an adjustable suture would circumnavigate this issue.

Some surgeons have advocated for essentially transposing the entirety of both vertical rectus muscles without disinserting them. After a small resection of the lateral halves of the vertical rectus muscles, the muscle bellies are transposed anterolaterally toward the insertion of the lateral rectus muscle. They are then attached to the sclera along the spiral of Tillaux using an adjustable suture (Figure 1F).[12] Proponents of the technique argue that it redirects the force of the entire vertical rectus muscle as opposed to just part of it (in contrast to split-muscle techniques), permits adjustment, and spares the anterior ciliary arteries of all of the rectus muscles. Published data are insufficient to document the relative efficacy of this approach. Furthermore, as described,[12] the small transverse incision in each vertical rectus muscle just anterior to where the suture is placed could still endanger the anterior ciliary supply.

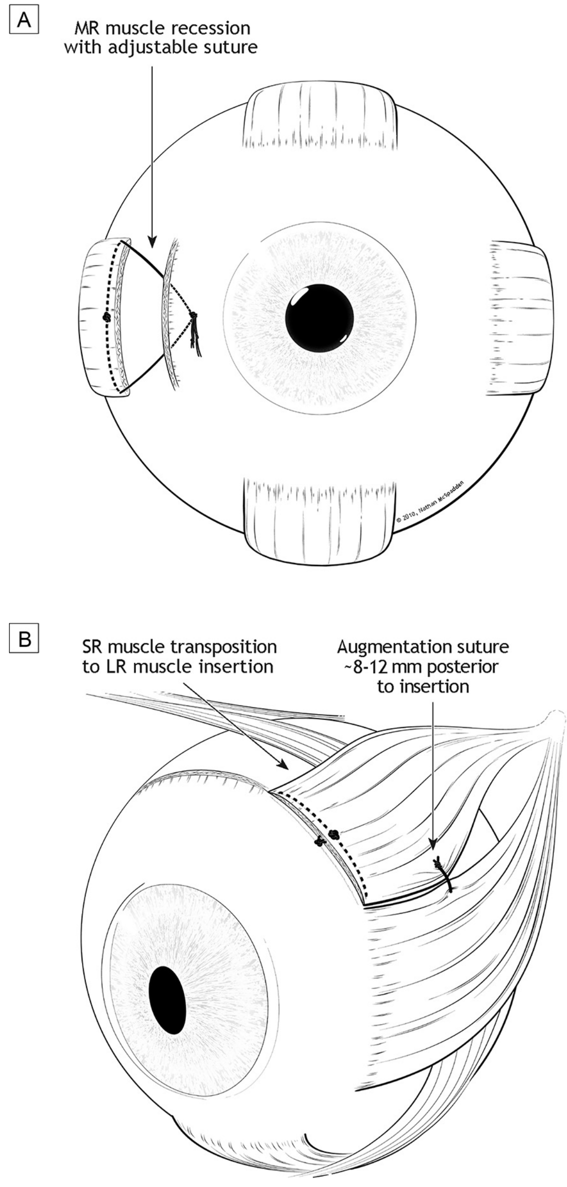

FIG 3. Diagrammatic representation of a combined superior rectus transposition (SRT) and medial rectus recession procedure with adjustable suture. A, The medial rectus recession on adjustable suture. B, The SRT procedure with augmentation suture. Although the amount of muscle included in the augmentation suture should be one-quarter to one-third the width of the muscle belly, it may appear that the augmentation suture includes less than one-quarter of the muscle width after tying the knot. This technique spares both anterior ciliary arteries of the inferior rectus muscle. Reproduced with permission from Mehendale and colleagues.[36]

Superior Rectus Transposition

Others have proposed transposing just one vertical rectus muscle to spare some of the ciliary circulation and to simplify the procedure. SRT involves transposing only the superior rectus muscle to the insertion of the lateral rectus muscle along the spiral of Tillaux, leaving the inferior rectus muscle untouched, usually in combination with ipsilateral medial rectus recession, with or without an augmentation suture (Figure 3). A review of the outcomes of this approach in patients with esotropic DS and complete loss of lateral rectus muscle function showed significant improvement in the mean angle of esotropia, abduction limitation, and head turn, with recovery of stereopsis in 80% of patients who did not have it preoperatively.[36] No clinically significant induced vertical or torsional diplopia was observed; 1 of 19 patients had an overcorrection that required further surgery.[3] This closely parallels outcomes from the original description of the procedure (Johnston SC, et al. ARVO abstract; IOVS 2006; 47:ARVO E-Abstract 2475), which also showed an improvement in the BSV range of 15°-45°.

SRT Augmented by Ipsilateral Medial Rectus Weakening

Whether to perform SRT alone or in combination with ipsilateral medial rectus recession is determined by several factors. As with VRT, if the medial rectus muscle is tight, patients are more likely to not respond to transposition alone and to require combined (or subsequent) medial rectus recession.[13] Another factor is the magnitude of the initial deviation. SRT alone appears to be sufficient to correct an esotropia of up to 15^{Δ}.[36] When the esotropia is $> 15^{\Delta}$, combining SRT with an adjustable medial rectus recession of up to 5 mm is advisable,[36] based on the amount of esotropia in primary position and whether the medial rectus muscle is tight on forced ductions. For larger deviations (usually $\geq 50^{\Delta}$), SRT can be combined with bilateral adjustable medial rectus recessions.[22,25]

SRT Compared to Medial Rectus Weakening

Compared to medial rectus recession alone for esotropic DS, SRT, either in isolation or in combination with ipsilateral medial rectus recession, results in similar improvement in esodeviation and head turn. However, to achieve the same effect SRT requires a smaller medial rectus recession, which reduces the risk of a postoperative adduction deficit. Furthermore, a much higher percentage of patients undergoing SRT show an improvement in abduction of at least 1 unit compared with those undergoing non-SRT surgery (80% vs 30%). Finally, patients undergoing SRT experience recurrence of esotropia (and resultant head turn) less frequently.[3,37]

SRT Compared to VRT

In their retrospective comparison of VRT and SRT (with medial rectus recession) in patients with abducens nerve palsy, Lee and Lambert[38] found that SRT showed an equally beneficial effect on the esotropia, a greater improvement in abduction, and less need for further intervention (eg, medial rectus botulinum toxin injection or recession). It also showed no greater risk of inducing new, persistent, induced vertical deviations or torsional diplopia.[38] It remains to be determined whether these findings are applicable to DS; nevertheless, SRT appears to offer several advantages over VRT for esotropic DS.

The degree of abduction achieved with SRT alone appears to be comparable to that achieved with VRT alone (imputed across different studies; there are no prospective, comparative trials), despite requiring only half the surgery. This has obvious time and cost implications. When combined with medial rectus recession, SRT may result in greater improvement in abduction than VRT alone, despite both being two-muscle surgeries. There may also be less risk of ASI, not only for isolated SRT but perhaps even when combined with medial rectus recession, because the vertical rectus muscles are thought to contribute more blood flow to the anterior segment.[39] By combining SRT with medial rectus recession on an adjustable suture, the horizontal and vertical alignment can be fine-tuned in the postoperative period; this can be particularly useful in DS, where surgery (especially transpositions) can yield unexpected results. Finally, simultaneous medial rectus recession can also help address any tightness of the medial rectus muscle that risks causing residual or recurrent esotropia after the procedure.

SRT Complications

One concern with SRT is the risk of causing significant vertical misalignment or unwanted torsional effects due to a theoretically imbalanced transposition. In practice, however, the data do not support these concerns.[3,36] In fact, the SRT complication rate compares favorably to the VRT data.[22-24] Although asymptomatic hypotropia in abduction has been noted in several cases of DS after SRT, careful review revealed that the hypotropia was in fact present prior to surgery in all cases.[40] Regardless, it seems advisable to place the transposed muscle on an adjustable suture in both children and adults.[28,29] If an induced vertical deviation does occur with SRT, it is usually a hypertropia from an overactive or tight superior rectus muscle, which can often be identified by preoperative assessment or by intraoperative forced ductions and ameliorated by recessing the muscle up to 3 mm at the time of the transposition.

Another theoretical risk of SRT is the creation of postoperative torsional imbalance, because lateral transposition of the superior rectus muscle may enhance its intorsional effect. The data in esotropic DS again do not appear to support this concern.[3,36] There appears to be little to no induced intorsion following SRT (even with augmentation suture placement), whether measured subjectively by double Maddox rods or objectively by direct fundus visualization.[36,41] There is a single reported case of a patient who developed 10° of induced intorsion following SRT, although the same patient also showed by far the greatest horizontal overcorrection in the study; thus, this case may be atypical.[42] It is unclear why vertical and torsional complications do not occur more frequently after SRT, although there are several theories.[3,36] Although the concerns about vertical and torsional deviations have not been borne out in practice, either may occur, and so it would seem advisable to counsel patients about these risks.

Inferior Rectus Transposition

IRT is a logical alternative to SRT. When combined with an augmentation suture, either with or without simultaneous adjustable medial rectus recession, IRT was shown to significantly improve the esotropia, abduction, and head turn in a small group of patients who were deemed at risk of hypothetical exacerbation by SRT due to preoperative hypertropia or intorsion of the operative eye.[43] IRT can also achieve a small downward shift in the operative eye that can help correct a preexisting hypertropia. The initial report was in the context of abducens nerve palsy, so a role for primary IRT in DS is yet to be defined.

Secondary IRT (ie, following prior SRT, which effectively makes it a staged VRT) can be performed to augment the effect of the initial SRT surgery. One early report of an isolated case of IRT following SRT suggested that the magnitude of incremental benefit of this approach was modest.[36] However, a more recent report documented 5 patients with abducens nerve palsy who underwent initial SRT with medial rectus recession, for supramaximal esotropia of 80^{Δ}-100^{Δ} and abduction deficit of −4 to −7, but were left with a significant residual esotropia (30^{Δ}-60^{Δ}) and abduction deficit (−3 to −4). Their residual esotropia was successfully eliminated with secondary IRT.[44] In this series, SRT alone or in combination with medial rectus recession achieved adequate correction in 4 patients with a similar degree of preoperative esotropia (80^{Δ}-95^{Δ}). However, on average their abduction deficit was less (−3 to −4), as was their duration of esotropia (0.5-2 years vs 1.5-22 years), suggesting that these variables should be considered during any surgical planning. It remains to be determined whether such findings hold for DS.

Unilateral Exotropic DS

Exotropic DS occurs in about 10%-15% of cases. It usually presents with a tight lateral rectus muscle caused by marked anomalous innervation, resulting in exotropia in primary gaze, as well as exotropia, globe retraction, and up- and downshoots in adduction. Nontransposition procedures often correct the exotropia in primary position and improve the head turn, although the globe retraction and up- and downshoots frequently persist.[2] Hence some surgeons advocate for periosteal fixation or extirpation of the lateral rectus muscle in exotropic DS,[13] either in isolation or in combination with a transposition procedure to help return some abducting ability to the eye once the lateral rectus muscle function has been eliminated.[45]

Lateral Rectus Disinsertion with or without Partial VRT for Exotropic DS

Lateral rectus disinsertion and periosteal fixation alone appears to achieve a greater improvement in the exotropia and adduction deficit in exotropic DS when performed without concomitant partial VRT (unsurprisingly, because there is no antagonistic abducting force).[46] By contrast, simultaneous partial VRT restores some abducting ability to the eye, leading to a greater range of BSV (twofold) and a reduced risk of consecutive esotropia (0% vs 30%).[46] The two approaches seem to achieve a similar improvement in globe retraction in adduction (about 50%-60%). To our knowledge, there is no published data on the use of SRT or IRT after lateral rectus neutralization for exotropic DS. However, extrapolating the aforementioned VRT data,[46] the potential benefits of augmenting abduction using temporal SRT while sparing the inferior rectus muscle may be useful.

Lateral Rectus Disinsertion and Partial VRT for Esotropic DS

Not all patients with severe lateral rectus muscle co-contraction are exotropic in primary gaze. They can be esotropic but still require lateral rectus muscle neutralization to achieve the desired surgical outcome or to reduce up- and downshoots. Lateral rectus disinsertion and partial VRT for esotropic DS with severe co-contraction can improve the esotropia, head turn, abduction and adduction ability, and range of BSV in these patients.[13] However, some patients do require ipsilateral medial rectus recession 6 months or more after the initial surgery for recurrent esotropia in primary position,[13] raising concern for ASI, depending on the clinical scenario. Lateral rectus muscle disinsertion with simultaneous temporal SRT could be considered as a primary procedure in such cases to reduce the risk of ASI, should a delayed medial rectus recession be required. Combined medial and lateral rectus recessions with SRT in patients with symptomatic globe retraction who are deemed to be at low risk for ASI have also been performed.[47]

Nasal SRT for Exotropic DS

A nasal transposition procedure can be used to augment the adducting ability of the affected eye. One group reported good results of nasal SRT used to treat 2 patients with exotropic DS. One patient presented with orthotropia in primary position after prior surgical intervention, and the other was exotropic in primary position; both patients had clinically significant underaction of the medial and lateral rectus muscles as well as globe retraction on forced ductions. They underwent nasal SRT, with or without superior rectus recession (Schneider JL, et al. IOVS 2010; 51:

ARVO E-Abstract 3012). Based on limited data, the authors concluded that this approach can improve both eye motility and range of BSV in such patients. The aforementioned report on SRT by Mehendale and colleagues[36] excluded 1 adult patient with exotropic DS who underwent augmented nasal SRT along the Spiral of Tillaux to the insertion of the medial rectus muscle. However, this had to be surgically reversed 3 days later owing to intolerable extorsional diplopia. Hence, it remains to be determined if this is a viable surgical strategy for exotropic DS.

Bilateral Esotropic and Exotropic DS

Finally, 10%-15% of DS patients have bilateral disease,[48,49] with roughly half being esotropic, one-third being orthotropic, and the rest being exotropic.[49] In terms of management, bilateral DS has been categorized according to the presence or absence of fusion.[4] Those with fusion often have limited eye movements, minimal head turn, and unpredictable surgical outcomes; nonsurgical approaches (eg, prisms) are often advisable in these cases.[4] Those without fusion can present with marked esotropia or exotropia, depending on the amount of anomalous innervation to the lateral rectus muscle in each eye and which eye is used for fixation. In such cases, the desired surgical outcome can often be achieved by balancing the abducting and adducting forces in the dominant eye.[4] Although unilateral surgery may suffice, a bilateral approach is sometimes necessary to balance the forces between the two eyes.[50]

In bilateral esotropic DS cases, the deviation in primary position can be as great as 70^{Δ}.[51] Bilateral medial rectus recessions in these patients reduce the adducting ability of the operative eye.[2] The surgical dose depends on the magnitude of deviation in primary gaze, the range of ductions, and the amount of restriction on forced ductions. Some patients also require abduction augmentation; VRT can be useful in this scenario.[52] Britt and colleagues,[53] reporting the results of a large case series, concluded that VRT can significantly improve esotropia in primary position at both distance and near as well as the abducting ability of the operated eye(s), and can aid in the acquisition of fusion in some cases. Patients with an esotropia of $<25^{\Delta}$ underwent unilateral VRT, whereas those with esotropia of $>25^{\Delta}$ underwent bilateral VRT. All patients undergoing bilateral VRT, and some undergoing unilateral VRT, also received augmentation sutures. Only 2 of 11 patients developed mild adduction limitation, and 1 required further surgery for an induced vertical deviation.[53]

SRT outcomes in bilateral esotropic DS have also been reported. Bilateral SRT, followed by later unilateral 3.5 mm medial rectus recession, significantly improved the primary position esotropia, head turn and abduction limitation in 1 patient with bilateral esotropic DS.[54] In contrast, unilateral SRT and concomitant ipsilateral medial rectus recession in a 10-month-old girl with bilateral esotropic DS (mentioned earlier in the context of torsional outcomes for SRT) showed a large horizontal overcorrection and intorsion that required surgical reversal.[42] She was very hyperopic (+13.5 to +15.0 D), although full refractive correction was worn for the preoperative evaluation. The authors noted the similarity to another reported case: a 1-year old girl with unilateral DS who showed significant horizontal overcorrection following SRT with ipsilateral medial rectus recession[36]; her deviation did not improve until the SRT had been reversed, the medial rectus muscle advanced to its original insertion, and the lateral rectus muscle recessed 12 mm. Age may be a determinant, because it is also a known risk factor for overcorrection after VRT.[22] Some advise against surgery for DS in patients <1 year of age unless there is a clear threat to fusion.

Bilateral exotropic DS is the least common form reported. It can be associated with type 1, 2, or 3 DS and even with different types in each eye.[55] Surgical planning in these cases must be tailored to a given clinical scenario. We are not aware of any reported transposition procedure outcomes for this form of DS.

Finally, there are several important features to consider when planning surgery for bilateral DS. A key point is that one is always measuring a secondary deviation, as the contralateral (fixating) eye also has paretic or restricted horizontal rectus muscles. Therefore, one must be cautious about generating too great an effect with any planned surgery, which risks overcorrection (and argues for the use of adjustable sutures in these patients). A further consideration is whether there is a pattern to the deviation. Some bilateral esotropic DS patients demonstrate a V pattern, although A patterns have also been observed.[56] Surgical correction should address any observed pattern in addition to the horizontal deviation.

Two important considerations for bilateral transposition surgery for bilateral DS (and other causes of bilateral esotropia) are the potential effects of induced vertical and torsional deviations. On the one hand, a bilateral transposition procedure (VRT, SRT, or IRT) that induces a vertical deviation of similar direction and magnitude in each eye should not create vertical diplopia. In contrast, if the transposition induces a small-to-modest torsional effect that a patient can typically fuse when performed unilaterally, bilateral surgery can precipitate a torsional diplopia, because the effect will be additive between the two eyes. So far this has not been reported, at least in the context of bilateral SRT with bilateral medial rectus recession for bilateral abducens nerve palsy related to either trauma or Moebius syndrome.[57,58]

Conclusions

Over the last 50 years, transposition procedures have continued to evolve, expanding the repertoire of interventions available to strabismus surgeons for the treatment of all types of DS. These powerful procedures should be used routinely but always with caution, given the complex and variable presentation of DS and the unpredictability of outcomes even with seemingly simple interventions.

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