



# An Insight of Intermittent Exotropia Management and Its Complications

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## Abstract

**Background:** Intermittent exotropia (XT) is an exodeviation controlled by fusional mechanisms and usually preceded by exophoria. Unlike phoria, intermittent XT spontaneously breaks into manifest XT. Intermittent XT is a common form of childhood exotropia, accounting for about 50%-90% of all the exotropia and affecting about 1% of the general population. Once intermittent XT becomes manifest, it either remains unchanged or gradually deteriorates. Rarely, it undergoes partial disappearance. Intermittent XT is usually first observed by the parents in early childhood as a spontaneous drifting out of one eye mostly when the child is tired, sick or day-dreaming. Symptoms of intermittent XT include blurred vision, asthenopia, visual fatigue, and rarely diplopia in older children and adults. The main goal of treatment in intermittent XT is to preserve the binocular vision. Several surgical approaches have been used successfully to treat exotropia. Bilateral lateral rectus recession (BLRR), unilateral lateral rectus recession and unilateral medial rectus resection, as well as bilateral medial rectus resection (BMRR) have all been used to treat this condition. The choice of procedure classically has been based on the measured distance/near incomitance. In Lateral rectus muscle recession; By moving a rectus muscle posterior to its original insertion site and reattaching it to the sclera, the length/tension curve of the muscle is changed. It produces the effect of "weakening" the muscle's effect on the globe. For most recessions, this effect is seen clinically only as a change in the alignment of the eye. This weakening effect probably occurs due to reduction in the distance between the origin and new insertion of the muscle, and changes in the relationship between Tenon's capsule, the rectus muscle pulleys and the intermuscular septum.

**Keywords:** Intermittent Exotropia Management, Lateral rectus muscle recession

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## Introduction

Intermittent exotropia (XT) is an exodeviation controlled by fusional mechanisms and usually preceded by exophoria. Unlike phoria, intermittent XT spontaneously breaks into manifest XT. [1]

Intermittent XT is a common form of childhood exotropia, accounting for about 50%- 90% of all the exotropia and affecting about 1% of the general population. Once intermittent XT becomes manifest, it either remains unchanged or gradually deteriorates. Rarely, it undergoes partial disappearance. [2]

Intermittent XT can be classified according to the disparity of distance and near deviation, **Burian's classification**:

- 1) *Basic type*; defined as the distance deviation equals or less than 10 prism diopters (PD) of near.
- 2) *True divergence excess type*; defined as exodeviation at distance 10 PD larger than at near fixation, with no post-occlusion change in the deviation at near.
- 3) *The pseudo-divergence excess type*, defined as exodeviation at distance 10 PD larger than at near fixation, but with increased near deviation within 10 PD when monocular occlusion is applied
- 4) *The convergence insufficiency type*, defined as greater 10 PD or more exodeviation at near than at distance. [3]

Exodeviations are classified by near measurement according to:

- A. Short-duration monocular occlusion to exclude fusional stimuli.
- B. Adding +3.00 D spheres removes the effect of accommodative convergence. [4]

## Presentation and Diagnosis

Intermittent XT is usually first observed by the parents in early childhood as a spontaneous drifting out of one eye mostly when the child is tired, sick or day-dreaming. The tropic phase tends to increase in degree, frequency and duration. At a later stage, the phoric phase may be retained only for near (tenacious proximal fusion) and the patient can be constantly exotropic at distance using suppression or abnormal retinal correspondence during most of his/ her waking hours. [5]

Symptoms of intermittent XT include blurred vision, asthenopia, visual fatigue, and rarely diplopia in older children and adults. Many patients with intermittent exotropia have photophobia (squinting to bright light) that was originally thought to be a way for eliminating diplopia or confusion. [6]



The age of onset of intermittent XT coincides with the age of visual maturation for children, which is between 3 and 6 years. As a rule, during the phoric phase of intermittent XT, the eyes are well aligned, and the patient may have a bifoveal fixation with excellent stereopsis ranging between 40 and 60 arc second. During the tropic phase, most patients will show large regional suppression of the temporal retina and abnormal retinal correspondence. [7]

A minority of patients with intermittent XT may have the monofixation syndrome and do not develop normal bifoveal fixation or stereopsis and may even have a significant amblyopia.[8]

#### **Measuring the exodeviation:**

For diagnosis of exotropia, a complete eye examination is conducted including assessment of ocular motility and measurement of ocular deviation for gaze at near (33 cm.) and distance (6 m).

Alternate prism cover test is used to measure the maximum deviation. This test is performed at distance and near while the patient views an accommodative target in the primary position and wearing his refractive correction. Base in prism is placed over one eye and dissociation continued by alternate cover of the both eyes simultaneously, with gradually increasing prism strength, until there is no movement (the highest prism power that induced no movement before reversal of the deviation).[9].

#### **Treatment of intermittent exotropia:**

##### **A. Non-surgical treatment:**

1. Correction of refractive errors
2. Treatment of amblyopia if present.
3. Part time occlusion of non-deviating eye.
4. Base out prism (exercising prisms) to stimulate accommodative convergence.

##### **B. Surgical treatment:**

##### **⇒ *Indications of surgery***

The main goal of treatment in intermittent XT is to preserve the binocular vision

1. Poor/worsening of control (Newcastle Control Score).
2. Increasing angle of deviation.
3. Decrease in stereopsis for distance or near.
4. Double vision.
5. Parental demand.
6. Cosmotoc.[10]

Several surgical approaches have been used successfully to treat exotropia. Bilateral lateral rectus recession (BLRR), unilateral lateral rectus recession and unilateral medial rectus resection, as well as bilateral medial rectus resection (BMRR) have all been used to treat this condition. The choice of procedure classically has been based

on the measured distance/near incomitance.[11].

Early temporary postoperative overcorrection up to 10–20 Prism Diopter (PD) has been recommended in surgical treatment of intermittent XT due to tendency towards postoperative exotropic drifts then functional results are more stable. Initial consecutive ET with small angle disappears spontaneously with time but larger angles tend to increase.[12].

### **Limbus-insertion distance**

#### **Definition:**

Limbus-insertion distance (LID) of lateral rectus (LR) is the distance between the grey-white line of the corneal limbus and the midpoint of the posterior edge of the LR muscle insertion and measured about 6.9mm.[13]

#### **Clinical significance**

There is positive correlation between LID and dose response effect. When LID was <5.5 mm, the dose response was smaller and when LID > 6.25, dose response is larger, so increasing amount of recession is recommended in smaller LID and decreasing amount of recession in large LID to achieve best results. [13].

### **Lateral rectus muscle recession**

By moving a rectus muscle posterior to its original insertion site and reattaching it to the sclera, the length/tension curve of the muscle is changed. It produces the effect of “weakening” the muscle’s effect on the globe. For most recessions, this effect is

seen clinically only as a change in the alignment of the eye. This weakening effect probably occurs due to reduction in the distance between the original and new insertion of the muscle, and changes in the relationship between Tenon’s capsule, the rectus muscle pulleys and the intermuscular septum.(14)

Amount of surgery used in bilateral LR muscle recession in patients with basic XT (mm). (14).

Prior to surgery a decision is made as to the type of anesthesia to be performed. Options for anesthesia include general, retrobulbar or peribulbar block, sub-Tenon’s injections, or topical. Considerations for anesthesia selection include the laterality of surgery, age of the patient, and associated medical conditions. (14)

#### **Pre-operative measures**

Once in the operating room, the periorbital skin is sterilized with povidone-iodine or hexachlorophene, after which the ocular surface is rinsed with half-strength povidone-iodine solution. Draping of the face and head is performed to maintain sterility.(14)

#### **Isolating the muscle**

A lid speculum is used to retract the lids and to keep the eyelashes out of the surgical field. Forced ductions are performed to identify any restrictions or limitations. The first incision through conjunctiva is made either in the fornix or at the limbus. A limbal incision may provide better exposure and is thought to cause less scarring because Tenon’s

capsule is not as traumatized.(15) However, a limbal incision is thought to affect the anterior segment blood supply more and can be more uncomfortable post-operatively. Fornix incisions have the advantage of better cosmetic appeal post-operatively with less risk to the anterior segment circulation. Dissection is performed to bare sclera, through the conjunctiva, Tenon's capsule and the intermuscular septum. A Stevens or similar hook is then passed between Tenon's and sclera, with the plane of the hook parallel to the insertion of the muscle. Traction is placed on the hook towards the limbus to isolate the fibers of the muscle insertion. A larger muscle hook, such as a Jameson, Helveston or Greene hook, is then passed behind the Stevens hook to ensure all muscle fibers are isolated. The insertion of the muscle is then exposed by removing any Tenon's attachments if performing a limbal incision or by freeing up and retracting the conjunctiva and draping it over the toe of the hook if using a fornix incision. The intermuscular septum at the toe of the hook is incised and special attention is made to ensure all muscle fibers are obtained with a pole test. The muscle is carefully cleaned of all its fascial attachments and check ligaments.(14)

### Recession

- The anterior insertion of the muscle is cleaned and adequately exposed.
- A double armed 6-0 suture on a spatulated needle is used to secure the muscle insertion. Absorbable sutures such as Vicryl (Ethicon) are commonly used. The muscle is secured in a manner to facilitate a recession via scleral suturing or a hang-back technique.
- For scleral suturing, a partial thickness muscle pass exiting at the muscle margin is made followed by a full thickness single or double locking bite at both the superior and inferior poles.
- For the hang-back technique, a central locking bite is made first, followed by the procedure described above. The muscle is disinserted from the globe by placing traction on the muscle hook and the sutures to keep them away from the insertion.
- Westcott scissors are then used to disinsert the muscle from the sclera. The cut stump of the insertion remaining on the sclera is grasped with forceps at both the superior and poles to maintain orientation of the globe.
- For scleral suturing, calipers set at the predetermined amount of recession are used to mark the distance posterior from the poles of the original insertion for placement of the muscle.
- The posterior sclera is marked to correspond with both the superior and inferior aspects of the insertion. The needles attached to the previously disinserted muscle are passed partial thickness through the sclera with visualization of the needle through the superficial sclera lamellae.
- The direction of the pass is angled slightly anterior to parallel with the insertion. Special caution is taken to ensure that a perforation or

penetration of the globe does not occur. This is repeated with the second arm of the suture which is passed at the second marked site posterior to the insertion.

- The scleral exit sites of the needles usually are closely approximated. The “crossed sword” technique is thought to allow for a more secure knot. The sutures are then pulled anteriorly to bring the muscle up to the new recessed position on the sclera. The sutures are then tied and trimmed. For the hang-back technique, after the muscle has been disinserted, both arms of the suture are passed through half-thickness sclera at the original insertion and pulled up to the insertion.
- The sutures are clamped with a locking needle driver and calipers are used to measure the number of millimeters for the intended recession from the exit of the scleral tunnels. The sutures are securely tied with 3 square knots and cut. The muscle is then allowed to retract in a hang-back fashion and the amount of recession is verified with calipers.(16).

Complications of lateral rectus recession

Intraoperative Surgical Site Complications

### Scleral Perforation

Perforations may occur in the normally thinned sclera directly behind the muscle insertion, as well as during the dissection, isolation, and disinsertion

of the muscle tendon. The reported incidence of scleral perforation varies widely with current estimates ranging from 0.3% to 7.8%. (17)

Risk factors increased if the sclera is thinned, such as in high myopia with a staphyloma, and if there is significant scarring or hemorrhage (more common during a re-operation) that lead to poor exposure and visualization of the sclera for suture placement. (17)

To decrease risk of perforation, using a magnified view of the surgical field and placing flexible spatulated needles through the sclera with good exposure and clear visibility of the needle tip at all times.(18)

Sclera perforation can lead to significant other problems such as endophthalmitis, vitreous hemorrhage, or retinal detachment. (18)

It is managed by laser retinopexy which may be required at the time of perforation if a large retinal tear or a small retinal tear in a patient at high risk for retinal detachment is noted. Laser retinopexy may not be necessary for a child with well-formed vitreous at low risk for detachment.(18)

### Lost Muscle

A lost muscle occurs when the extraocular muscle slips free of the sutures or surgical instruments during surgery with no direct attachment between the muscle tendon and the globe, allowing the muscle and its



capsule to both retract posteriorly into the orbit. **(18)**

Lost muscle can be minimized by using gentle surgical techniques when isolating and securing the muscle belly and rotating the globe towards a resected muscle rather than pulling the tight muscle anteriorly. The risk can also be reduced by direct placement of scleral sutures at the site of the new insertion, instead of utilizing a hang-back technique from the original insertion.**(19)**

It is diagnosed post operatively on extraocular motility testing and the patient may present with a large angle, incomitant strabismus. Imaging can also help localize lost muscles.

Management: For a lost muscle, an attempt should be made to retrieve the muscle promptly, during the same surgery if possible Coats and Olitsky, 2007.. If the muscle cannot be retrieved, a transposition surgery can be considered, although there is a risk of anterior segment ischemia by performing surgery on three eye muscles (the lost muscle plus two transposed muscles) at the same time. Depending on the location, a posterior orbital approach with the aid of an orbital surgeon can sometimes successfully isolate and retrieve the lost muscle.**(19)**

### **BCSC Pediatric Ophthalmology and Strabismus.**

A slipped muscle occurs when less than full-thickness sutures are used to capture the muscle tendon, effectively only capturing the superficial muscular capsule instead of securing the muscle belly**(18)**. Post-operatively, the muscle

belly retracts within the muscle capsule when force is exerted during contraction, leading to clinical weakness of the operated muscle. **(20)**

The risk factors are similar to those for a lost muscle. The risk of a slipped muscle increases when the muscle is tight, such as with dysthyroid orbitopathy or the contracted antagonist of a paretic muscle. It can be difficult to create space at the muscle insertion to allow a full-thickness suture pass to encompass both the muscle capsule and muscle belly. **(18)**

Primary prevention: The risk of a slipped muscle may be minimized by using full-thickness bites when passing the muscle suture. Special groove hooks or muscle clamps can be used with tight muscles to provide more space to securely place the muscle sutures to encompass the muscle belly in addition to the muscle capsule. Some physicians prefer to use two locking bites instead of the traditional one locking bite to ensure adequate capture of the muscle fibers.**(16)**

It is diagnosed by limitation of ocular movement post operatively but less than lost muscle.**(20)**

A slipped muscle can be repaired during surgery by locating the muscle capsule attached to the globe from the prior surgery. By following the muscle capsule posteriorly, the muscle itself can be found, isolated, and attached to the globe.**(16)**

### **Oculocardiac Reflex**

The oculocardiac reflex (OCR) is a vagal bradycardic response to compression of the eyeball and/or tension on an extraocular muscle tendon, which stimulates the trigeminal nerve. **(21)**

It is managed by stopping the triggering stimulus will terminate the reflex. Pharmacologic management includes intravenous administration of anticholinergics, such as atropine. **(22)**

Most infections occur around the initial surgical incision into the conjunctiva and present within the first week after. Rarely, infections can penetrate deeper into the orbit with proptosis, eyelid swelling, chemosis, and erythema in the classical presentation of orbital cellulitis. Sometimes endophthalmitis can develop, either with or without a scleral perforation. The most commonly cultured organisms include *Staphylococcus aureus* (MRSA and MSSA), group A *Streptococcus*, and coagulase-negative *Staphylococcus*. One study reported an overall incidence of post-operative infection of 0.14%**(23)**, while estimates of the incidence of endophthalmitis range from 1 in 350,000 cases to 1 in 18,500 cases<sup>1</sup>.**(18)**

The risk of post-operative infection may be minimized by aggressively treating any superficial infection or bacterial overgrowth pre-operatively, by using meticulous sterile technique during surgery, and by using post-operative antibiotics.

Signs of infection include conjunctival injection, eyelid erythema and swelling, discharge, eye pain and/or increased tenderness over the surgical site, systemic signs, fever, and photophobia. Patients with severe

infections tend to present with systemic infections. **(23)**

Orbital imaging with magnetic resonance imaging (MRI) or computed tomography (CT) can help identify preseptal and orbital cellulitis. For infections, gram stains and cultures (either of the conjunctiva or, in the case of scleral perforation and possible endophthalmitis, of the vitreous) can aid with diagnosis and treatment.**(18;23)**

Management: For infections, topical antibiotics are used to treat conjunctivitis, systemic antibiotics are used to treat preseptal and orbital cellulitis, and intravitreal antibiotics are used to treat endophthalmitis. The medical follow up is determined by the severity of the clinical problem. For serious infections, the patient may require inpatient hospitalization for intravenous antibiotics and close monitoring. For less serious problems, the follow up period can be longer, with specific instructions to call for any apparent deterioration in clinical status. If imaging demonstrates a subconjunctival or orbital abscess, then abscess drainage be necessary**(18;23)**

### **Pediatric Ophthalmology and Strabismus.**

A foreign body granuloma can develop sporadically in susceptible patients, usually a few weeks after surgery**(24)**. Pyogenic granuloma, also known as lobular capillary hemangioma, occurs with proliferation of capillaries with edema typically at the conjunctival incision. Pyogenic granuloma can occur after any incision or trauma to the conjunctiva. **(24)**

The risk of a foreign body granuloma may be reduced by avoiding gut sutures and by proper draping to keep lashes out of the surgical field**(18)**.

Persistent post-operative foreign body sensation can be caused by a foreign body granuloma. Examination will demonstrate a localized, elevated, hyperemic mass at the suture site typically less than 1 cm in diameter.**(18)**

Regarding treatment, Topical corticosteroids are used for several weeks, with possible surgical excision if no clinical response is observed. **(25)**

### **Conjunctival Inclusion Cyst**

A conjunctival inclusion cyst can occur when conjunctival epithelial cells are buried beneath the conjunctival surface during surgery **(18)**. These cells can multiply over time to create a subconjunctival cyst days to years after the original surgery. The incidence of conjunctival inclusion cysts range from 0.25% to 0.4%**(26)**

### **Conjunctival Scarring**

Conjunctival scarring occurs when, instead of returning to the typical translucent white appearance, the conjunctiva remains chronically hyperemic and pink, particularly after a second or third operation<sup>[21]</sup>. This complication can be exacerbated by advancement of Tenon's capsule too close to the limbus, particularly during a resection, or by advancement of the plica semilunaris onto the bulbar conjunctiva. If severe, the conjunctival scarring itself can cause a restrictive strabismus.**(16)**

The risk of conjunctival scarring is increased for a re-operation and after a surgical resection. Care must be taken to avoid creation of conjunctival foreshortening or symblepharon. The risk of conjunctival scarring may be minimized by careful wound closure. If persistent, it may require surgery to remove scar tissue and smooth the conjunctival surface. The risk of excessive conjunctival scarring increases with each surgery performed. A visible conjunctival scar is present in over 90% of cases postoperatively and is usually subtle and clinically insignificant, although it can rarely be severe. **(27)**

### **Dellen**

Dellen are shallow, clearly defined excavations at the margin of the cornea or sclera. They occur when thickened bulbar conjunctiva (either from scarring, hemorrhage, or swelling) prevents adequate and even lubrication of the ocular surface during a blink. Any disruption of the tear layer on the sclera or cornea causing local dehydration can create a dellen.**(16)**

The risk of dellen formation is higher for a limbal incision (6.5%) than a non-limbal (2.2%) incision, because the subsequent irregularity of the perilimbal conjunctiva can cause a disruption of the tear layer in the anterior sclera and cornea<sup>[16]</sup>. Dellen are more common after resection surgery than recession surgery.**(16)**

### **Anterior Segment Ischemia**

Anterior segment ischemia occurs when blood supply to the anterior segment through the ciliary arteries within the four rectus muscles is



interrupted(28). Simultaneous surgery on three rectus muscles in the same eye, or two rectus muscles in a patient with compromised blood flow from vascular disease, can cause ischemia. The incidence of significant anterior segment ischemia has previously been estimated to be approximately 1 in 13,000 cases, although this is likely an underestimate because mild cases probably occur without clinical detection.(29)

### Change in Refraction

Change in the refraction post-operatively occurs from a change in the force the extraocular muscle places on the cornea through its attachment to the sclera. Over time, this change in force usually reaches a new equilibrium, typically with restoration of the original corneal refractive shape.(18)

### Diplopia

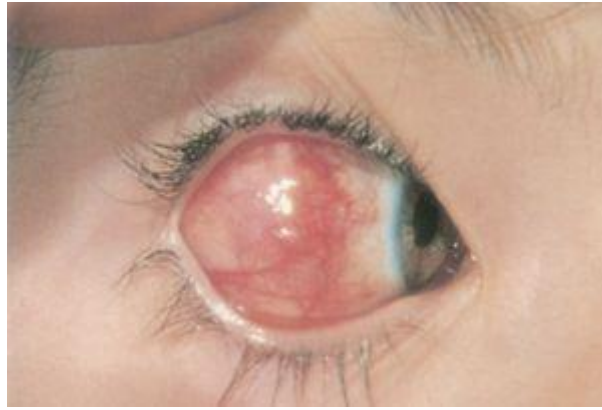
Diplopia can occur in patients capable of vision in each eye from an imperfect eye alignment.



**Figure 1:**Orbital cellulitis of the right eye 2 days after bilateral lateral rectus recession. **Image credit: BCSC Pediatric Ophthalmology and Strabismus.**

**Rutsteinetal1996.**Diplopia is more likely in adult patients who possess limited ability to suppress the second image. The risk is also higher in more complicated types of strabismus, particularly paretic forms of strabismus.(30;31)

One study found that 9% of adult patients had temporary diplopia after surgery, which typically resolved within 6 weeks. Only 0.8% developed persistent intractable diplopia. Postoperative diplopia, both temporary and intractable, is more likely in adults who experienced diplopia with preoperative prism testing(30).



**Figure 2:**Allergic reaction to chronic gut suture following strabismus surgery. Image credit: BCSC Pediatric Ophthalmology and Strabismus.



**Figure 3:**Pyogenic granuloma following left lateral rectus muscle surgery. Image credit: Espinoza GM, Lueder GT. Conjunctival pyogenic granulomas after strabismus surgery.

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