

# insight

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### **Sankara Nethralaya – The Temple of the Eye.**

It was in 1976 when addressing a group of doctors, His Holiness Sri Jayendra Saraswathi, the Sankaracharya of the Kanchi Kamakoti Peetam spoke of the need to create a hospital with a missionary spirit. His words marked the beginning of a long journey to do God's own work. On the command of His Holiness, **Dr. Sengamedu Srinivasa Badrinath**, along with a group of philanthropists founded a charitable not-for-profit eye hospital.

Sankara Nethralaya today has grown into a super specialty institution for ophthalmic care and receives patients from all over the country and abroad. It has gained international excellence and is acclaimed for its quality care and compassion. The Sankara Nethralaya family today has over 1400 individuals with one vision – to propagate the Nethralaya philosophy; the place of our work is an Alaya and Work will be our worship, which we shall do with sincerity, dedication and utmost love with a missionary spirit.

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## The era of scholars is here

Dr. S. Meenakshi

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**Correspondence:**

Dr. S. Meenakshi,  
Director, Academics,  
Senior Consultant,  
Department of Paediatric  
Ophthalmology & Strabismus



Do postgraduates need to be involved in research? Do they need to present papers and posters in conferences? Do they need to publish? These questions are often answered differently by different people, accrediting agencies and organizations. These activities have been studied under the loose title of 'scholarly activity'. Many accrediting bodies both in India and other countries are increasingly recommending that postgraduate training programs maintain an environment of inquiry and research. This is in addition to the thesis completion during the 3-year program that is considered an important research activity and is a requirement. Creating an environment for scholarly activity also includes providing faculty who have research interests, access to interesting patients, resources to scour the literature.

Studies have shown that postgraduate residents' participation in research was associated with higher levels of satisfaction about the residency program.<sup>1</sup> In a questionnaire based study of residents regarding such research activities, most of the respondents reported that their scholarly project was a worthwhile experience despite considerable barriers. They also added that teaching research skills more explicitly, having a focused curriculum to teach such skills and also including protected research time to their training program may add to the success of such activities during residency training.<sup>2</sup>

When one looks at the recommendations of accrediting bodies like the ACGME, the newer thinking among educational planners on the need for incorporating research into the curriculum becomes apparent. ACGME's stance on scholarly activity as per the 2016 document is as follows: "The curriculum must advance residents' knowledge of the basic principles of research, including how research is conducted, evaluated, explained to patients, and applied to patient care. Residents should participate in scholarly activity. The sponsoring institution and program should allocate adequate educational resources to facilitate resident involvement in scholarly activities."<sup>3</sup>

This issue of *Insight* showcases the observational, analytical and the scholarly activities of postgraduates of the CU Shah Postgraduate Ophthalmic Centre. It attempts to cover many subspecialties of ophthalmology and makes for interesting reading. I applaud the Editor for his vision and wish the postgraduates many years of inquiry and research.

### References

1. Takahashi O, Ohde S, Jacobs JL, et al *J Gen Intern Med* 2009;24:716.
2. Rivera JA, Levine RB, Wright SM. *J Gen Intern Med* 2005;20:366.
3. [https://www.acgme.org/Portals/0/PDFs/Specialty-specific%20Requirement%20Topics/DIO-Scholarly\\_Activity\\_Resident-Fellow.pdf](https://www.acgme.org/Portals/0/PDFs/Specialty-specific%20Requirement%20Topics/DIO-Scholarly_Activity_Resident-Fellow.pdf)

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# Type II Duane's retraction syndrome with severe upshoot with ipsilateral superior oblique muscle palsy: a rare presentation

Dr. Nimisha Sharma, Dr. Manideepa Banerjee, and Dr. S Meenakshi

## Correspondence:

Dr. Nimisha Sharma,  
Medical Research Foundation,  
Sankara Nethralaya,  
18 College Road,  
Nungambakkam, Chennai, India.  
Email: id:nimisha0310@gmail.  
com

## Abstract

Type II Duane's retraction syndrome (DRS) is the least common strabismus characterized by limitation of adduction with the presence of upshoot, downshoot or both. Abduction can be normal or slightly altered. Secondary muscle changes like fibrosis, and anomalous insertions have been associated with DRS. Its associations with dissociated vertical deviations have been reported. We report a rare case of DRS with superior oblique palsy presenting post trauma in a tertiary care centre.

## Introduction

Duane's retraction syndrome (DRS) is a unusual form of strabismus characterized by limitation of horizontal movements and globe retraction with palpebral fissure narrowing on attempted adduction of the affected eye.<sup>1</sup> In general, 1–4% of strabismic patients have Duane's syndrome.<sup>2, 3</sup>

Type II DRS is least common and presents as limitation of adduction with exotropia of the affected eye. Abduction can be normal or slightly affected. A characteristic upshoot, downshoot or both may occur in adduction.

Its association with dissociated vertical deviation has been reported previously.<sup>4</sup> Superior rectus overaction/contracture syndrome (SRSy) was described by Jampolsky in 1964. Superior rectus contracture has been previously reported in a patient with unilateral superior oblique palsy (SO).<sup>5</sup> Here, we report a rare case of type II DRS with ipsilateral SO.

## Case Report

A 50-year-old male presented with complaints of blurring of vision and double vision in downgaze following head trauma 6 days back, which was associated with black eye. There were no preceding systemic illnesses, and patient's birth history, family history and medical history were not significant. There was no history of surgical intervention in the past. Previous CT scan showed thickening of right superior oblique muscle.

On examination, best corrected visual acuity was 6/6 with no significant refractive error. The patient had small left head tilt with left face turn. Ocular motility showed global limitation in adduction with upshoot on adduction with palpebral fissure narrowing in the right eye. The alternate

prism cover test showed 20PD right hypertropia (RHT) with 6PD exotropia in primary gaze and 12 PD RHT with 6PD exotropia in downgaze. Deviation of near showed 14RHT with 6PD exotropia. Rest movements in both eyes were full. The patient had diplopia in a distance Worth 4-dot test. Double Maddox rod showed 10° extorsion in primary gaze and 15° in downgaze (Figs 1 and 2).

Diplopia and torsion were noted in diplopia charting and superior oblique underaction on Hess charting. Fundus examination showed mild extorsion. Review of the old photographs prior to trauma did not reveal any face turn. There was a diagnostic dilemma regarding the co-existence of SO with DRS. Although right hypertropia pointed towards the severe upshoot in adduction, the appearance of left face turn after trauma, and



Figure 1: Left face turn.



Figure 2: Extraocular movements in all gazes showing global limitation in adduction with upshoot on adduction with palpebral fissure narrowing in the right eye.

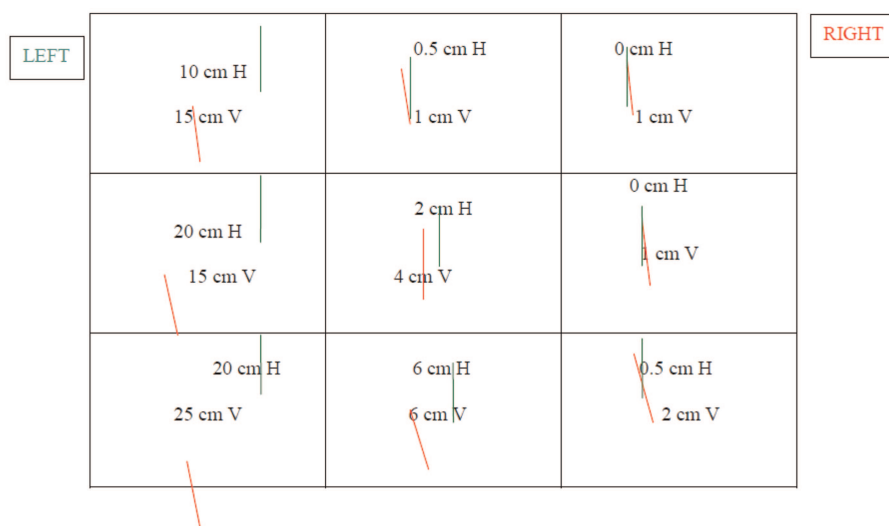
large vertical and torsional diplopia in the field of action of superior oblique muscle and underaction of the same in Hess charting, suggested the presence of right SO. The presence of exotropia, adduction limitation with upshoot and palpebral fissure changes, on the other hand, confirmed diagnosis of Type II DRS. Subsequently, surgery was planned to correct his face turn and vertical squint. On table, inferior oblique muscle was found to have an abnormal insertion. Forced traction test showed laxity in the right superior oblique muscle. A superior oblique tuck of 6 mm with inferior oblique recession in the right eye was performed. A repeat traction test at the conclusion showed no evidence of iatrogenic Brown's. On the first postoperative day, his face turn and small head tilt improved significantly with collapse of the vertical squint to a large extent. He was kept

under close follow-up and the next plan of action is to take care of the exotropia and the residual upshoot by lateral rectus recession with or without a Y-split (Figs 3 and 4).

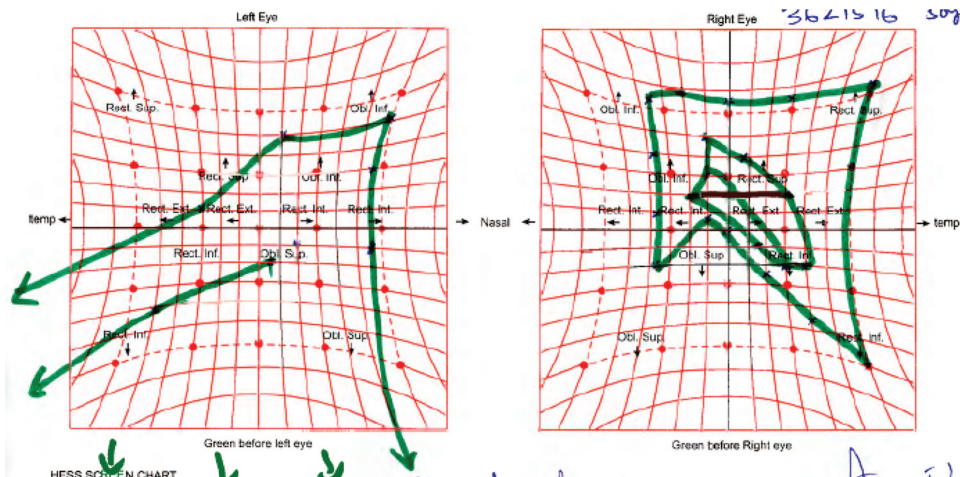
**Discussion**

Heuck described patients with severe limitation of ocular motility and retraction of the globe in nineteenth century.<sup>6</sup>

Type II DRS is the least common type (<10%) characterized by a marked limitation of adduction with exotropia of the affected eye, abduction normal or slightly limited, retraction of the globe and narrowing of the fissure on attempted adduction. On electromyography, the lateral rectus showed peak impulses on abduction and a second paradoxical peak on attempted adduction. There was normal behaviour of the medial rectus.



**Figure 3:** Diplopia chart showing increased separation of the vertical images in the field of action of superior oblique muscle and the presence of torsional diplopia more in downgaze.



**Figure 4:** HESS chart showing severe underaction of superior oblique muscle and mild overaction of inferior oblique muscle in the right eye and overaction of superior oblique and inferior rectus along with underaction of superior rectus in the left eye.

The pathogenesis of DRS has a wide spectrum, which can be classified into mechanical, anatomical and innervational disorder. Anatomical changes most commonly seen are fibrotic changes and anomalous insertions.<sup>7</sup> Mechanical disturbances can occur due to the presence of facial bands found in some cases of DRS. These band causes the limitation of eye movement.

An upshoot may occur due to co-innervation of superior rectus muscle with the lateral rectus or could be because of the mechanical factors as the bridle or leash effect due to tight lateral rectus.<sup>3</sup>

The innervational type of upshoot is characterized by the presence of hypertropia in the primary position.<sup>8</sup> Another characteristic feature is that, in innervational type, there is a gradually increasing upshoot of the eye as it moves in adduction. Our case had the obvious features of exotropic DRS with a limitation of adduction, upshoot and palpebral fissure narrowing in adduction.

In our case, the exact pathogenesis of SO is not known. The laxity of the superior oblique tendon on forced traction testing hint towards a possibility of congenital SO. But the absence of significant head tilt even prior to trauma and sudden onset vertical and torsional diplopia in the field of action of superior oblique muscle following trauma points more towards an acquired SO with DRS.

### Conclusion

DRS and SO have been shown to co-exist in our case although the exact pathogenesis still remains

unravelling. Identification of each of these components requires a meticulous history taking, review of old photographs and thorough clinical examination and supportive investigations. The plan of surgery in such cases is a staged procedure: first, correcting the SO by superior oblique tucking with inferior oblique recession and, second, DRS surgery, as recession of lateral rectus muscle on the involved side in proportion to the size of exotropia<sup>9</sup> with or without Y split for upshoot.

### References

1. DeRespinis PA, Caputo AR, Wanger RS, Guo S. Duane's retraction syndrome. *Surv Ophthalmol* 1993;38:257-88.
2. Ahluwalia BK, Gupta NC, Goel SR, Khurana AK. Study of Duane's retraction syndrome. *Acta Ophthalmol (Copenh)* 1988;66(6):728-30.
3. Kirkham TH. Inheritance of Duane's syndrome. *Br J Ophthalmol* 1970;54(5):323-9.
4. Khawam E, Ghazi N, Salti H. "Jampolsky Syndrome". *Superior rectus overaction-contraction syndrome: prevalence, characteristics, etiology and management. Binocul Vis Strabismus Q* 2000;15:331-42.
5. Scott AB, Wong GY. Duane's syndrome. *An electromyographic study. Arch Ophthalmol* 1972;87:140-7.
6. Hueck G. Über Angeborenenvererbten Beweglichkeitsdefect der Augen. *Klin Monatsbl Augenheilkd* 1879;17:253.
7. Gobin MH. Surgical management of Duane's syndrome. *Br J Ophthalmol* 1974;58:301-6.
8. Kraft SP. Surgical approach to Duane syndrome. *J Pediatr Ophthalmol Strabismus* 1988;25:119-30.
9. Rosenbaum AL. Costenbader lecture. The efficacy of rectus muscle transposition surgery in Duane syndrome and VI nerve palsy. *J AAPOS* 2004;8(5):409-19.

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# A case of non-resolving subconjunctival cysticercosis managed by surgical excision

Chanda Gupta and Md. Shahid Alam

## Correspondence:

Md Shahid Alam,  
Associate Consultant,  
Department of Oculoplasty,  
Sankara Nethralaya,  
Email: mshahidalsm@gmail.com

## Introduction

Cysticercosis is an infestation caused by the dissemination of the larval form *Cysticercus cellulosae* of *Taenia solium* (pork tapeworm) or rarely *Taenia saginata* (beef tapeworm). The most common form of systemic involvement is neurocysticercosis.<sup>1</sup> Ocular and adnexal cysticercosis represents 13% to 46% of systemic disease.<sup>2</sup>

The most common site of localization reported in India is orbit and adnexa, whereas in Western world it is in the vitreous cavity or the sub-retinal space.<sup>3</sup> Among the orbital cysticercosis, extraocular muscle (EOM) involvement is the commonest.

Incidence of subconjunctival cysticercous cysts among the cystic lesions of conjunctiva is 8.89%.<sup>6</sup>

We report here a case of subconjunctival cysticercosis which was managed by surgical excision.

## Case Report

A 16-year-old student from West Bengal presented to us with complaints of swelling in the upper part of the right eye which was gradual in onset and constant in size since 3 months associated with pain. There was no significant past ocular or systemic history. The patient had undergone computed tomography (CT) scan of the brain and orbit 1 month before elsewhere (Fig. 1).

On examination, the best corrected visual acuity was 6/5, N6 in both the eyes. On examination, the right upper lid, a small 1 × 1 cm firm

and mobile mass was at the 12 o'clock meridian near the insertion of the superior rectus muscle.

The left eye was within normal limits. Ocular movements were full and free. There was no proptosis and retrobulbar resistance was normal. Anterior segment and posterior segment were normal in both eyes. Our radiologist opined that the CT scan showed a small cyst with the scolex in the superior periocular region anterior to the insertion of the superior rectus muscle in the tendon of the levator palpebral superioris muscle, suggestive of cysticercosis.

Conservative treatment was started after obtaining physical fitness from our physician. She was prescribed oral wysolone 1 mg per kg body weight per day in tapering doses and oral albendazole 15 mg per kg body weight per day for 6 weeks. At 6 weeks, there was no change in the size of the lesion.

She underwent excision biopsy of the mass under general anaesthesia. She was started on topical steroids, lubricants and analgesics post-operatively (Fig. 2).

Histopathological examination showed a cyst from the epibulbar region; the lumen contained oral and intestinal parts of the tapeworm. There were numerous giant cells, epitheloid cells, lymphocytes and plasma cells suggestive of cysticercosis, capsulated with surrounding granulomatous inflammation. On the basis of the history, clinical examination and investigations, a diagnosis of subconjunctival cysticercosis of the right eye was made (Fig. 3).



Figure 1:

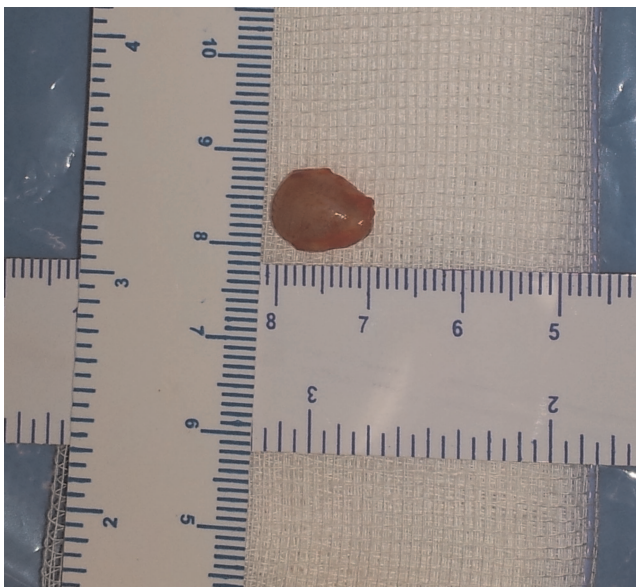


Figure 2:

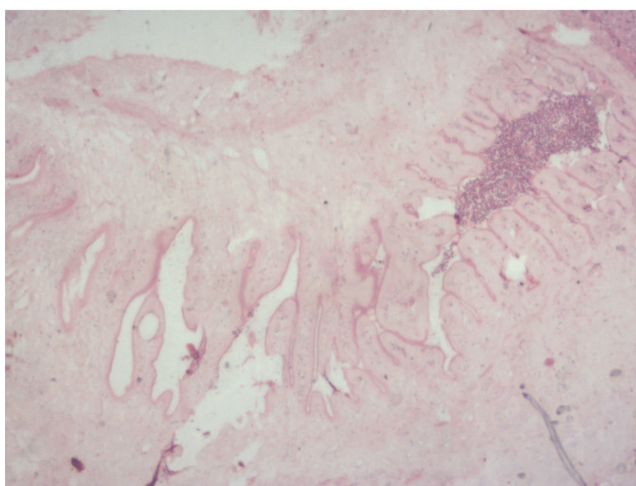


Figure 3:



## Discussion

Cysticercosis is an infestation caused by *Cysticercus cellulosae*, the larval form of the cestode *Taenia solium*.<sup>7</sup> On infestation of the undercooked pork which contains cysticerci of *T. solium*, the larval cyst is released and the inverted scolex attaches to the host intestine. The adult worm grows and releases proglottids in the stool which contaminates the environment. Thus, humans play the role of a definitive host. Cattle ingests the proglottids and releases thousands of eggs. The eggs develop into oncosphere that becomes a cysticercus in the intermediate host. So, when humans ingest undercooked pork, they develop taeniasis (infection with an adult worm), whereas when they ingest eggs, they suffer from cysticercosis (infection with the larva). Pigs are the definitive hosts and humans play a role of intermediate host. Infection with eggs results through infected food handlers or by ingestion of fruit and vegetables fertilized with contaminated human waste and by contaminated water supplies in endemic areas.<sup>6, 8</sup> The eggs develop into a cysticercus in the small intestine and reach other tissues via the haematogenous route.

The patient can present with floaters, moving sensations, pain, redness, photophobia, diminution of vision, seizures and meningitis (Neurocysticercosis). On rupture of the cyst, the patient may develop anterior uveitis, vitritis, rhegmatogenous or exudative retinal detachment, disc oedema and phthisis bulbi.<sup>9</sup> The cysticercosis subconjunctival lesions tend to present as hyperaemic epibulbar masses that are sometimes fluctuant.<sup>10</sup>

Diagnosis of cysticercosis is usually made on history, clinical examination and radiological investigations like magnetic resonance imaging (MRI) and CT scan and by histopathology when it can be excised. B scan ultrasonography is useful in detecting the scolex.

Medical therapy with albendazole and oral steroid is recommended for the EOM form and orbital cysticercosis.<sup>11</sup>

In our case, the patient was not responding to the medical treatment, so we went ahead with the surgical excision.

The patient is doing well without any recurrence of cysticercosis.

## References

1. Grover AK, Puri P. Orbital myocysticercosis presenting as subconjunctival abscess. *Ind J Ophthalmol* 1996;44:229–31.
2. Mais FA. Cryosurgery in ocular cysticercosis. [in Portuguese]. *Rev Bras Ophthalmol* 1969;28:99–106.
3. Pushker N, Bajaj MS, Betharia SM. Orbital and adnexal cysticercosis. *Clin Exp Ophthalmol* 2002;30:322–33.
4. Cano MR. Ocular cysticercosis. In: Ryan SJ, Glaser BM, Michels RG, editors. *Retina*. 2nd ed. St. Louis: CV Mosby; 1994, p1553–8.
5. Madigubba S, Vishwanath K, Reddy G, Vemuganti GK. Changing trends in ocular cysticercosis over two decades: an analysis of 118 surgically excised cysts. *Indian J Med Microbiol* 2007;25(3):214–219.
6. Rath S, Honavar SG, Naik M, Anand R, Agarwal B, Krishnaiah S, Sekhar GC. Orbital cysticercosis: clinical manifestations, diagnosis, management, and outcome. *Ophthalmology* 2010;117:600–605.
7. Sharma T, Sinha S, Shah N, Gopal L, Shanmugam MP, Bhende P, Bhende M, Shetty NS, Agrawal R, Deshpande D, Biswas J, Sukumar B. Intraocular cysticercosis: clinical characteristics and visual outcome after vitreoretinal surgery. *Ophthalmology* 2003;110:996–1004.
8. Prasad KN, Prasad A, Verma A, Singh AK. Human cysticercosis and Indian scenario: a review. *J Biosci* 2008;33(4): 571–82.
9. Nath K, Gogi R, Zaidi N, Johri A. Cystic lesions of conjunctiva (a clinicopathological study). *Indian J Ophthalmol* 1983;31(1): 1–4.
10. Singh A, Devendra J, Garg C, Dokania A. Subconjunctival cysticercosis: a case report. *IJSS Case Reports Rev* 2015;2(4): 11–14.
11. Pushker N, Bajaj MS, Chandra M, Neena . Ocular and orbital cysticercosis. *Acta Ophthalmol Scand* 2001;79:408–13.

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# Monocular elevation deficiency: when the involved eye takes up fixation

Prabrisha Banerjee and Meenakshi Swaminathan

## Correspondence:

Prabrisha Banerjee,  
Medical Research Foundation  
Sankara Nethralaya  
18, College Road,  
Chennai - 600 006  
email: prabrit.prabrisha@gmail.com

## Introduction

Monocular elevation deficiency (MED) is a form of restrictive strabismus, characterized by the unilateral limitation of elevation and associated ptosis.<sup>1</sup> The inability of the eye to elevate persists across the horizontal plane, from adduction to abduction. It is usually a sporadic condition.<sup>2</sup> This condition was previously described by Dunlap<sup>3</sup> as Double Elevator Palsy and the limitation of elevation was attributed to the paralysis of both the superior rectus and the inferior oblique muscle. MED can be congenital or acquired. Congenital MED is often seen in identical twins and it is caused by either of the following<sup>4</sup>: (a) paralysis of the superior rectus muscle without any involvement of the inferior oblique muscle. (b) Primary inferior rectus restriction. Metz found inferior rectus restriction without elevator weakness in 73% of MED patients.<sup>5</sup> (c) Lesion in the supranuclear pathway of upgaze located in the pretectum. (d) Inferior rectus restriction secondary to long-standing superior rectus palsy.

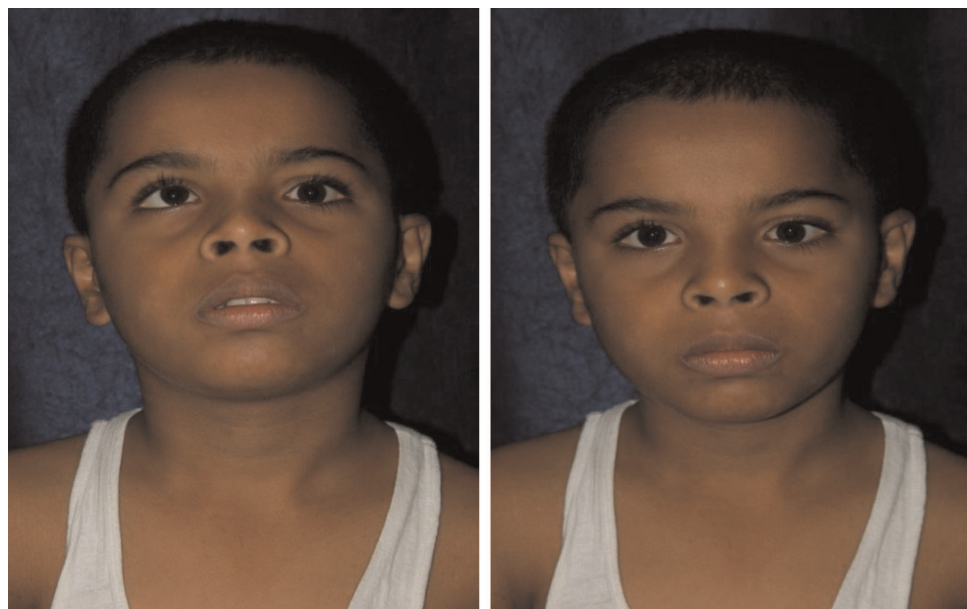
Acquired MED is seen in (a) cerebrovascular diseases, (b) tumours of the mid-brain, (c) syphilis, (d) sarcoidosis.<sup>1-4</sup> Congenital MED presents with (i) the inability to elevate the eye above midline in abduction, adduction or from primary position of gaze.<sup>4</sup> (ii) Hypotropia of the affected eye on the affected side. However, one-third of the cases

present as orthophoria in primary gaze.<sup>2</sup> Rarely the affected eye may fixate. (iii) Ptosis of the affected eye or sometimes pseudoptosis may be seen. (iv) Chin-up position.

Important investigations include forced duction test, active force generation test, saccadic velocity measurements, and magnetic resonance imaging of brain.<sup>5</sup> Some other conditions may mimic MED such as Brown syndrome, thyroid eye disease, congenital fibrosis of inferior rectus muscle, orbital floor fracture and myasthenia gravis.<sup>4, 5</sup> The indications for surgical interventions are (a) large vertical deviation in primary gaze, (b) vertical deviation causing suppression and amblyopia, especially in children and (c) abnormal head posture.<sup>4</sup> Now we are going to describe a rare case of MED where the patient is fixating with the affected eye.

## Case Report

An 11-year-old male child presented with diminution of vision and upward deviation of right eye for 3 years. He had undergone a surgery for bronchial fistula at 7 years of age. He was a full-term baby with normal birth weight. There was no history of delayed developmental milestones. There was neither any history of ocular disorder in the family nor any history of consanguinity.



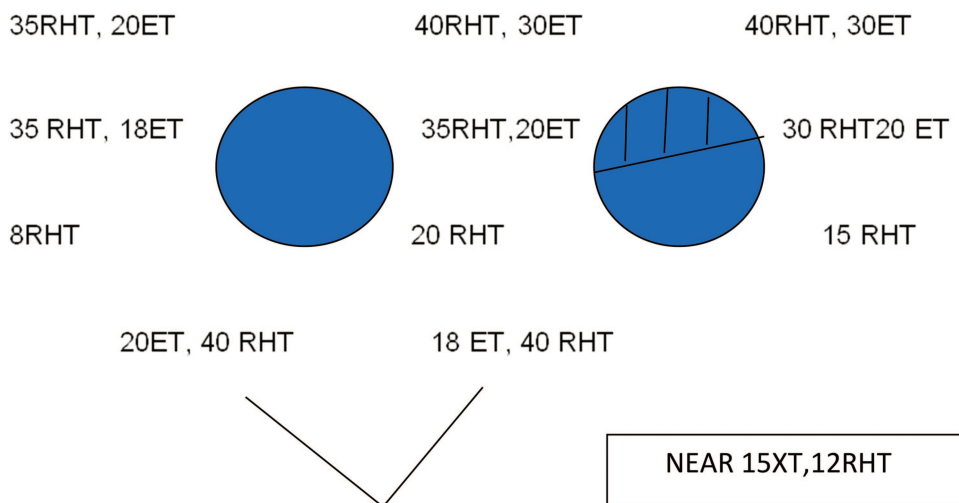
**Figure 1:** (a) Chin-up position. (b) Left eye taking up fixation.

Examination revealed visual acuity in the right eye to be 6/36 and in the left eye to be 6/6. He had a chin-up (~30°) position as shown in Figure 1(a). At primary position of gaze, the right eye was hypertropic and the left eye was hypotropic. Left eye was the fixating eye (Figure 1(b)). Ocular motility was full in the right eye, but there was marked limitation of elevation from primary position, in abduction and adduction in the left eye. There was globe retraction and unnatural convergence on attempted elevation. Mild to moderate ptosis was seen in the left eye which remained constant in all gaze (Figure 2). Fusional status was assessed by Worth 4-Dot test for distance as well as for near, and it revealed right eye suppression. Stereo acuity was about 140 s of arc. The alternate prism cover test showed 35RHT,

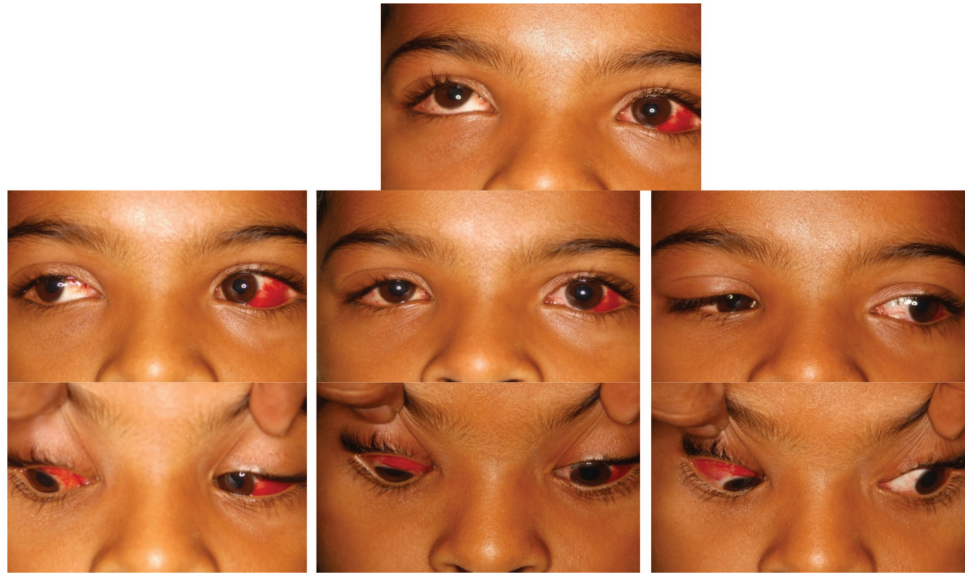
20ET at primary gaze, 40RHT, 30ET at upgaze, 20RHT at downgaze, 30RHT, 18ET at right gaze and 30RHT, 20ET at left gaze for distance and 12RHT, 15XT for near (Figure 3). Rotatory nystagmus was seen in both of the eyes. Pupillary reaction was normal. Slit lamp findings, intraocular pressure measured by Goldmann Applanation Tonometry and fundus findings of both of the eyes were within normal limit. MRI brain and orbit revealed cord-like thick inferior rectus muscle of left eye suggestive of fibrosis. FDT, performed preoperatively, was positive for left inferior rectus and right superior rectus under anaesthesia. Left inferior rectus recession (5 mm) and right eye superior rectus (3 mm) recession were performed under general anaesthesia. Postoperatively there was improvement in



**Figure 2:** Preoperative photos showing left eye hypotropia in primary gaze, limitation of elevation in upgaze from primary position, in adduction as well as in abduction.



**Figure 3:** Strabismus evaluation by alternate prism cover test.



**Figure 4:** Postoperative photo showing improvement in elevation and orthophoria in primary position.

elevation in the left eye as shown in Figure (4). He could fixate with both eyes. He was advised occlusion of left eye.

### Discussion

The child shows fixation preference for the involved eye, i.e. the left eye. When the hypotropic left eye fixates, there is an overshoot in the right eye. This is based on the Hering's law, where an excess innervation is required by the left superior rectus muscle to fixate leading to concomitant excess supply to the elevators of the right eye, thus causing hypertropia.<sup>5, 6</sup> The hypertropia in the right eye has been long standing which is justified by the chin-up position adapted by the child to facilitate fusion.<sup>5</sup> Eventually amblyopia has developed in the right eye leading to low vision. Ptosis is due to levator palpebrae superioris weakness. The persistence of ptosis when left eye is fixating rules out pseudoptosis which is seen in hypotropia due to the fascial attachments between the levator palpebrae superioris and superior rectus and disappears as the affected eye takes up fixation.<sup>4</sup> The fact that it is long standing has been reinforced by the MRI findings suggestive of inferior rectus fibrosis of the left eye. Preoperative FDT indicates restriction of left inferior rectus, which may be primary or secondary.<sup>5</sup> Surgical intervention is indicated as the child has already developed suppression and amblyopia. To align the hypertropic right eye in primary position weakening of right eye superior rectus is needed in addition to left eye inferior rectus recession.<sup>3, 7</sup>

Occlusion therapy should be continued and the child should be followed up regularly.

### Conclusion

A thorough ocular examination should be performed, including ocular motility, test for fusion and stereo acuity, cover, uncover and alternate prism cover test to reach a diagnosis. FDT holds importance in diagnosing the cause and management is based on the results of FDT. Early detection is important to prevent suppression and amblyopia of the normal eye.

### References

1. Von Noorden GK, Campos EC. Paralytic Strabismus. *Binocular Vision & Ocular Motility*, 6th edn. USA: Mosby, p442-443.
2. Kanski JJ, Bowling B. Strabismus. *Clinical Ophthalmology*, 7th edn. Elsevier, p774.
3. Bagheri A, Sahebghalam R, Abrishami M. Double elevator palsy, subtypes and outcomes of surgery. *J Ophthal Vis Res* 2008; 3(2):108-113.
4. Varshini S. *Monocular Elevation Deficiency*. Postgraduate Ophthalmology, 1st Edition. New Delhi: Jaypee, pp2064-2066.
5. Wilson ME, Wright KW. *Vertical Strabismus*. Pediatric Ophthalmology and Strabismus, 2nd edn. New York: Springer, 2003.
6. Khurana AK, Khurana I. *Extra Ocular Muscles and Ocular Motility*. Anatomy and Physiology of Eye, 2nd edn. New Delhi: CBS, pp285-326.
7. Bandyopadhyay R, Shetty S, Vijayalakshmi P. Surgical outcome in monocular elevation deficit: a retrospective interventional study. *Indian J Ophthalmol* 2008;56:127-133.
8. Kim JH, Hwang JM. Congenital monocular elevation deficiency. *Ophthalmology* 2009;116:580-584.

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## Isolated primary foveal hypoplasia

K. Gowri Pratinya, Chetan Rao, and Ramakrishnan Subramanian

### Correspondence:

K. Gowri Pratinya,  
Medical Research Foundation  
Sankara Nethralaya  
18, College Road,  
Chennai - 600 006  
email: pratinyak@gmail.com

### Introduction

Foveal hypoplasia is a morphological abnormality which represents different stages of arrested development of fovea. It encompasses complete absence (foveal aplasia) or partial absence (foveal dysgenesis) of foveal structure. It is characterized by the absence of foveal pigmentation and/or the foveal avascular zone. It may occur in isolation or in association with other conditions such as aniridia, albinism, achromatopsia, microphthalmos, retinopathy of prematurity (ROP), myopia and incontinentia pigmenti (IP).<sup>1-4</sup> The clinical diagnosis might often be missed due to the subtle nature of the fundus findings. This is a case report of an isolated foveal hypoplasia in which optical coherence tomography (OCT) was used to confirm the clinical suspicion of foveal hypoplasia in a 4-year-old boy.

### Case Summary

Mother of a 4-year-old boy was referred to the vitreous-retina service of our tertiary eye care hospital with the presenting complaints of child having shaking of the eyeballs since birth with abnormal head posture and with no history of any day or night blindness, photophobia, or premature birth. Parents have a history of second-degree consanguinity, and father of the child has nystagmus and microphthalmos. On examination he had a vision of OU - c/us/m with OD : CTC : +0.50 DS -0.75 DC 180 AXIS OS : CTC : -1.00 DS -0.50 DC 180 AXIS correction. He had a left exotropia with the right face turn and OU pendular nystagmus with normal pigmented iris. Systemic examination is unremarkable. Dilated fundus examination revealed normal optic nerve heads and absent foveal reflex with the vessels passing over the area of incipient fovea (Figures 1 and 2).

OCT was carried out, which showed the absence of foveal depression with the persistence of the inner retinal layers. The diagnosis of isolated primary foveal hypoplasia was made, and the patient was prescribed glasses and asked for a regular follow up (Figures 3 and 4).

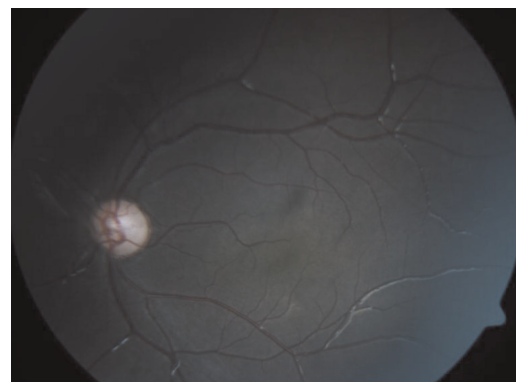
### Discussion

There were no signs of regressed ROP, myopia, cataract, corneal pannus, ocular albinism or IP as has been reported previously<sup>1-4</sup> in association with the foveal hypoplasia in our patient.

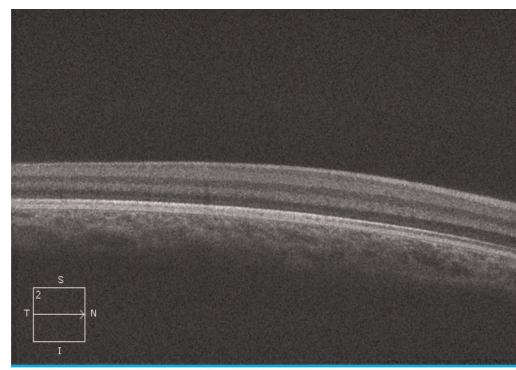
The role of OCT in the diagnosis of this case was also obvious. There was preservation of the inner retinal layers in the foveae of both the eyes as demonstrated by the OCT.



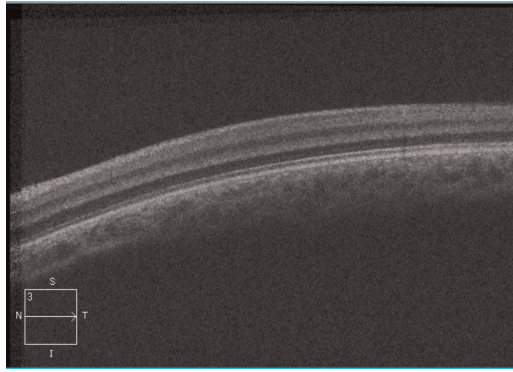
**Figure 1:** Fundus photograph of the right eye showing absent foveal reflex with the vessels passing over the area of incipient fovea.



**Figure 2:** Fundus photograph of the left eye showing absent foveal reflex with the vessels passing over the area of incipient fovea.



**Figure 3:** OCT of the right eye which showed the absence of foveal depression with the persistence of the inner retinal layers. Foveal thickness O: 260 µm.



**Figure 4:** OCT of the left eye which showed the absence of foveal depression with the persistence of the inner retinal layers. Foveal thickness OS 282  $\mu\text{m}$ .

This report illustrates the importance of careful examination of the foveal area in patients with unexplained visual loss with nystagmus that does

not fit into the other well-known dystrophies. The report also highlights the role of OCT in the diagnosis of this condition, by the demonstration of the preservation of the inner retinal layers in the fovea.

#### References

1. Oliver MD, Dotan SA, Chemke J, Abraham FA. Isolated foveal hypoplasia. *Br J Ophthalmol* 1987;71:926–930.
2. Recchia FM, Carvalho-Recchia CA, Trese MT. Optical coherence tomography in the diagnosis of foveal hypoplasia. *Arch Ophthalmol* 2002;120:1587–8.
3. Chen SD, Hanson R, Hundal K. Foveal hypoplasia and other ocular signs: a possible case of Incontinentia pigmenti? *Arch Ophthalmol* 2003;121:921.
4. O'Donnell FE Jr, Pappas HR. Autosomal dominant foveal hypoplasia and presenile cataracts: a new syndrome. *Arch Ophthalmol* 1982;100:279–81.
5. Hendrickson AE, Yuodelis C. The morphological development of the human fovea. *Ophthalmology* 1984;91:603–612.
6. Hendrickson A, Possin D, Vajzovic L, Toth C. Histological development of the human fovea from midgestation to maturity. *Am J Ophthalmol* 2012;154(5):767–778.e2.

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