AN OPHTHALMOLOGICAL STUDY OF ECTOPIA LENTIS, WITH MULTIFARIOUS AETIOPATHOGENESIS, DIVERSE CLINICAL PRESENTATIONS AND VARIED PROGNOSES

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ABSTRACT

BACKGROUND

Ectopia lentis is a hereditary or acquired displacement or malposition of the lens, from its normal position, with traumatic and non-traumatic aetiopathogenesis like homocystinuria due to cystathionine ß-synthase deficiency - chromosome 21q22 mutation, Marfan's syndrome; presenting with varying degrees of lenticular subluxation and manifesting near normal post-operative prognoses.

MATERIALS AND METHODS

It was a prospective study done on both the eyes of 4 patients (Cases A, B, C, D), which presented to Rajarajeswari Medical College and Hospital, Bangalore, with the complaints of diminution of vision. Case A also complained of progressive headache. Case B and case C gave history of past trauma. They were evaluated with detailed history of presentation and other significant past history. Detailed first visit examination was done, which included vision testing, anterior segment evaluation with slit lamp biomicroscopy and dilated fundus evaluation. They were admitted for further management. Relevant investigations and radiological investigations were done. Orthopaedic, ENT, Dental and Physician opinions were taken for systemic manifestations. Refractive corrections were done by manual small incision cataract surgery with anterior chamber intraocular lens or scleral fixated intraocular lens implantation or lensectomy with anterior vitrectomy and postoperative outcomes were recorded.

RESULTS

Case A showed 6/36 and 6/24 right and left eye vision, bilateral superonasal subluxation of lens with oval, sluggishly reactive pupil, early cataractous changes, iridodonesis and phacodonesis; with Marfanoid features and increased homocysteine levels. Case B showed visual acuity of 6/24 and counting fingers from 2 m in right and left eye respectively. Right eye was pseudophakic and left inferonasal subluxated lens, hypermature cataract, zonular dehiscence and phacodonesis. Case C showed right 6/60 and left 6/6 vision, with right inferonasally dislocated cataractous lens and phacodonesis. Case D showed right 6/60 and left 6/24 vision, bilateral inferiorly dislocated cataractous lens. After the surgical procedures, the patients recovered to almost near normal vision and ophthalmological conditions.

CONCLUSION

Case A was diagnosed as ectopia lentis due to homocystinuria, case B and case C as traumatic ectopia lentis and case D as idiopathic ectopia lentis. On proper refractive surgeries and further monitoring, all cases showed excellent post-operative prognoses and recovered well.

KEYWORDS

Ectopia Lentis, Homocystinuria, Traumatic Ectopia Lentis, Minimal Incision Cataract Surgery, Anterior Chamber Lens Implantation, Lensectomy with Anterior Vitrectomy, Scleral Fixation Intraocular Lens Implantation.

HOW TO CITE THIS ARTICLE: Hazra S, Hazra M. An ophthalmological study of ectopia lentis, with multifarious aetiopathogenesis, diverse clinical presentations and varied prognoses. J. Evid. Based Med. Healthc. 2018; 5(40), 2842-2850. DOI: 10.18410/jebmh/2018/583

Financial or Other, Competing Interest: None.
Submission 09-09-2018, Peer Review 13-09-2018,
Acceptance 26-09-2018, Published 29-09-2018.
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BACKGROUND

Berryat described the first reported case of lens dislocation in 1749, and Stellwag subsequently coined the term ectopia lentis in 1856.[1,2] Ectopia lentis is a hereditary or acquired displacement or malposition of the crystalline lens of the eye, from its normal position, due to the disruption or dysfunction of the lenticular zonular fibres containing high levels of cysteine; with either complete dislocation of the lens, rendering the eye functionally aphakic (luxation), or partial lenticular displacement, remaining partly within the pupillary area (subluxation).[3,4] Ectopia lentis can be

autosomal dominant (FBN1 polymorphism on chromosome 15q12), or autosomal recessive (ADAMTS17 mutation on chromosome 15q24) (with iris coloboma, aniridia, microspherophakia, ectopia pupillae).[5] Ectopia lentis becomes more marked with age and initiates glaucoma.[6] Three-quarters (approx..) of blind children in the world are in developing countries - Africa and Asia.[7,8]

Ectopia lentis is usually classified as following:

1. Traumatic (53%)
2. Non-Traumatic:
   - Genetic Ectopia lentis without systemic manifestation: 1. Simple ectopia lentis. 2. Ectopia lentis et pupillae
   - In Systemic disorders: Marfan's syndrome (superotemporal), Homocystinuria (inferotemporal), Weil- Marchesani syndrome, Hyperlysinaemia, Sulphite oxidase deficiency, Ehlers – Danlos syndrome, Crouzon disease, Refsum syndrome, Kniest syndrome, Mandibulofacial dysostosis, Sturge – Weber syndrome, Conradi syndrome, Pfaundler syndrome, Pierre syndrome, Wildervanck syndrome, Sprengel deformity.
   - In Ocular disorders: uveitis, hypermature cataract, pseudoexfoliation syndrome, ciliary body tumour, syphilis and trauma retinitis pigmentosa, persistant papillary membrane, aniridia, Reiger's anomaly, megalo cornea, blepharoptosis, high myopia, congenital glaucoma.[9]

MATERIALS AND METHODS

The study conducted was a prospective study. The duration of the study was 4 months, between September 2017 to December 2017. The examination was done on both the eyes of four patients with ectopia lentis, from the Ophthalmology Department of Rajarajeswari Medical College and Hospital, a tertiary care hospital. The inclusion criteria were as follows: (a) any age, of either sex; (b) patients with ectopia lentis; (c) willing for a post-operative follow-up; and (d) co-operative and conscious patients. The exclusion criteria were as follows: (a) uncooperative and unconscious patients; (b) patients presenting with acute severe/acute life-threatening/near-fatal clinical manifestations; (c) history of hypersensitivity or severe adverse effects to any of the study treatment; (d) pregnant or lactating women; (e) other associated medical illness having impact on study results; (f) patients not willing to participate in the study or adhere to the study protocol. The Institutional Ethical Committee approval and the written informed consent were obtained from all the study participants. After taking thorough clinical case history, general physical examination, vitals recording, systemic examination and ophthalmological examination were done and all the clinical findings were recorded. All the patients underwent a comprehensive preoperative evaluation of the anterior and posterior segments, including intraocular pressure measurements. A preoperative biometry was done, to assess the power of the intraocular lens which was to be implanted. All patients received preoperative antibiotic prophylaxis, started from one day prior to surgery. Any preoperative risk factor (e.g. poorly dilating pupil, pseudoexfoliation syndrome, intumescent cataract etc.) was recorded in every patient. Lensectomy with vitrectomy with spectacles correction or manual small incision cataract surgery with anterior chamber intraocular lens implantation or scleral fixation intraocular lens implantation with spectacles correction were performed, following the standard surgical protocol, under peribulbar anaesthesia by a single surgeon. The duration of the surgery, any intraoperative difficulties and complications were noted down.[10,11,12,13,14] All the patients received a standard regimen of topical antibiotics-steroid containing eyedrops, used every hourly for the first 7 days, and then every 2nd hourly for the next 7 days, and then gradually tapered over the next 4 weeks. The patients were reviewed on day 1, 7, 15, 30 and 45, and during each visit the best corrected visual acuity was recorded and a slit lamp evaluation, intra-ocular pressure assessment and fundus examination were done, and the post-operative prognoses were recorded.

RESULTS

Case	Age (Years)	History of Presenting Illness	Past History	Development History	Family History
A	10	1. Diminution of vision, insidious in onset, gradually progressive, painless, since 2 years, no aggravating and relieving factors 2. Headache 3. No history of trauma	1. Using spectacles, but uncomfortable 2. No history of stroke and epilepsy	No developmental delay	NS. No history of epilepsy in family
B	66	1. Diminution of vision in left eye, painless, sudden in onset, gradually progressive 2. History of trauma 1 week back	Cataract surgery of right eye, 2 years back	WNL	NS

C	7	1. Diminution of vision in right eye, sudden in onset, painless, gradually progressive 2. Diplopia since 1 week 3. No history of bleeding, watering or any aggravating or relieving factors 4. History of trauma to right eye with stick while playing, 1 week back	NS	WNL	NS
D	29	1. Diminution of vision more in right eye than in left eye, insidious in onset, gradually progressive, painless, since 3 months 2. No history of diplopia, trauma, watering and photophobia	NS	WNL	NS

Table 1. Details of Patient History

* Nothing significant – NS, within normal limits - WNL

Case	General Physical Examination	Systemic Examination	Visual Acuity	Slit-lamp Examination	Intraocular Pressure	Extraocular Movements	Colour Vision	Diplopia Charting	Fundo-scopy
a	weight: 32 kg, height: 131 cm, arm span: 139 cm	1. CVS, CNS, GIT, GIT: NAD 2. Orthopaedic examination: short stature, cubitus valgus, short fingers, increased carrying angle 3. Dental and ent examination: high arched palate, no crowding of teeth	Re: 6/36 p and LE: 6/24 p. improvement to RE: 6/24 p and Le: 6/18 p, with pinhole.	1. bilateral superonasal subluxation of lens 2. oval sluggishly reactive pupil bilaterally 3. bilateral early cataractous changes 4. zonules seen 5. bilateral iridodonesis and phacodones-is present 6. mild nystagmus present	RE: 16 mm hg and LE: 18 mm Hg	full and normal	normal	normal	WNL
b	WNL	1. CVS, CNS, GIT, RS: NAD 2. orthopaedic examination: WNL 3. Dental and ENT examination: WNL	RE: 6/24 p and le: counting fingers: 2 m. improvement to RE: 6/12 p and le: counting fingers: 2 m, with pinhole. near vision: N10.	RE: 1. pseudophakia, intraocular lens present in situ 2. pupil round, regular, reactive 3. clear cornea 4. ac quiet LE: 1. congested conjunctiva 2. inferonasal subluxated cataractous lens 3. hypermature cataract	RE: 16 mm hg and LE: 14.5 mm Hg	full and normal	normal	normal	RE: WNL and LE: hazy media due to hypermature cataract

				4. zonular dehiscence 5. phacodonesis 6. pupil: mild, dilated, sluggishly reactive 7. ac quiet					
c	WNL	1. CVS, CNS, GIT, RS: NAD 2. orthopaedic examination: WNL 3. dental and ENT examination: WNL	RE: 6/60 p and le: 6/6 p. improvement to re: 6/60 p and le: 6/6 p, with pinhole. near vision: n6.	RE: 1. cornea clear 2. subconjunctival haemorrhage present 3. mild dilated sluggish pupil 4. ac quiet 5. inferonasally dislocated lens 6. phacodonesis present 7. few vitreous strands seen LE: Anterior segment normal, pupil round, regular, reactive	RE: 16 mm hg and LE: 15 mm hg	full and normal	normal	diplopia present laterally more in superolateral quadrant	RE: hazy media and LE: WNL
d	WNL	1. CVS, CNS, git, RS: NAD 2. orthopaedic examination: WNL 3. dental and ENT examination: WNL	RE: 6/60 p and LE: 6/24 p. improvement to re: 6/36 p and le: 6/24 p, with pinhole.	1. bilateral lens dislocation inferiorly more in RE 2. both eyes cataractous, more in re 3. pupil round, regular, sluggishly reactive bilaterally 4. cornea clear 5. AC quiet 6. hypermature cataract in RE	RE: 14 mm hg and LE: 16.5 mm hg	Full and normal	Normal	Normal	RE: hazy media and narrowing of vessels and le: WNL

Table 2. Details of Examination

Cardiovascular system – CVS, Central nervous system – CNS, Gastrointestinal system – GIT, Respiratory system – RS, No abnormalities detected – NAD, Within normal limits – WNL, Right eye – RE, Left eye – LE, Anterior chamber – AC.

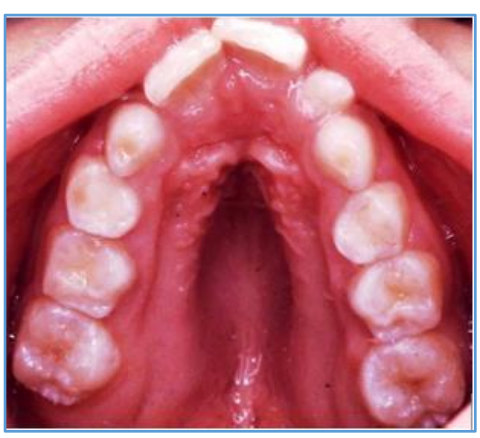

Figure 1. High Arched Palate in Case A: Ectopia Lentis Due to Homocystinuria with Clinical Features of Marfan's Syndrome

Case	Percentage of Lens Subluxation	Degree of Lens Subluxation
A	12%	Mild
B	53%	Severe
C	42%	Moderate
D	39%	Moderate

Table 3. Degree of Lens Subluxation

‡ Degree of lens subluxation classification:
Minimal to mild: 0% to 25%
Moderate: 25% to 50%
Severe: >50%

Case	Investigations Done	Investigation Report	Provisional Diagnosis
A	1. Homocysteine level 2. CBC 3. ESR 4. RFT 5. LFT 6. Serum electrolytes 7. RBS 8. ECG 9. Chest X-ray 10. MRI 11. CT Scan 12. VDRL 13. HIV 14. HBsAg	1. 39.14 mmol/L increased) 2. – 11. WNL 12. – 14. Negative	Ectopia lentis due to homocystinuria with clinical features of Marfan's syndrome
B	1. OCT 2. Bone Scan 3. X-ray Antero-posterior, Lateral 4. CBC with ESR 5. Serum electrolytes 6. RBS 7. ECG 8. RFT 9. LFT 10. MRI 11. CT Scan	1. – 11. WNL	Traumatic ectopia lentis
C	1. OCT 2. Bone Scan 3. X-ray Antero-posterior, Lateral 4. CBC with ESR 5. Serum electrolytes 6. RBS 7. ECG 8. RFT 9. LFT 10. MRI 11. CT Scan	1. – 11. WNL	Traumatic ectopia lentis
D	1. OCT 2. Bone Scan 3. X-ray Antero-posterior, Lateral 4. CBC 5. ESR 6. Serum electrolytes	1. – 14. WNL	Idiopathic ectopia lentis

	7. RBS 8. Serum homocysteine 9. Serum methionine 10. ECG 11. RFT 12. LFT 13. MRI 14. CT Scan		

Table 4. Investigations

OCT - Optical Coherence Tomography, CBC - Complete Blood Count, RBS – Random Blood Sugar, RFT – Renal Function Tests, LFT – Liver Function Tests, WNL – Within Normal Limits.

Case	Treatment	Post-Operative Treatment Visual Acuity									
		Day 1		Day 7		Day 15		Day 30		Day 45	
		Right Eye	Left Eye	RIGHT EYE	Left Eye	Right Eye	Left Eye	Right Eye	Left Eye	Right Eye	Left Eye
A	Lensectomy with anterior vitrectomy with spectacles (after 1 month) for both the eyes at 2 months interval	6/36 p	6/36 p	6/24 p	6/18 p	6/24 p	6/18 p	6/12 p	6/12 p	6/9 p	6/12 p
B	Manual small incision cataract surgery with anterior chamber intraocular lens implantation for the left eye at 3 months interval	6/24 p	Counting fingers: 2 m	6/24 p	6/60 p	6/24 p	6/36 p	6/24 p	6/24 p	6/24 p	6/24 p
C	Lensectomy with anterior vitrectomy and spectacles after 1 month for the right eye	Counting fingers: 2 m	6/6 p	6/60 p	6/6 p	6/18 p	6/6 p	6/9 p	6/6 p	6/9 p	6/6 p
D	Manual small incision cataract surgery with scleral fixation intraocular lens implantation and spectacles after 1 month for the right eye (Patient never returned for the review of the left eye)	6/60 p	6/24 p	6/60 p	6/24 p	6/36 p	6/24 p	6/36 p	6/24 p	6/36 p	6/24 p

Table 5a. Treatment and Postoperative Outcomes

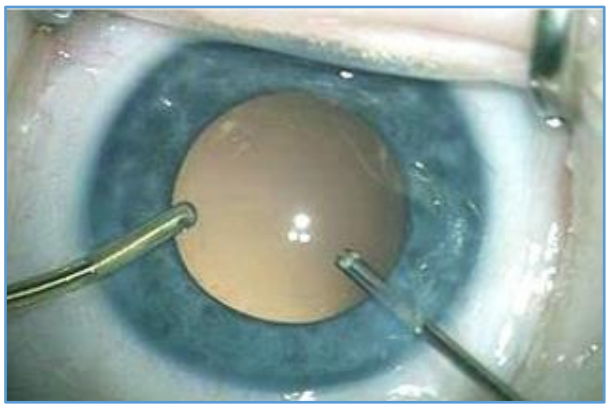

Figure 2. Lensectomy with Anterior Vitrectomy in Case A and Case C

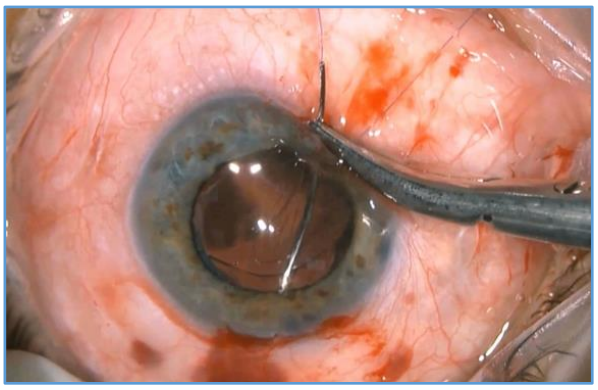

Figure 3. Scleral Fixation Intraocular Lens Implantation in Case D

Case		Post-Operative Treatment Slit-Lamp Examination					Post-Operative Treatment Intraocular Pressure (mm of hg)			
		DAY 1	DAY 7	DAY 15	DAY 30	DAY 45	DAY 1	DAY 7	DAY 15	DAY 30
A	RE	cornea hazy, pupil mid-dilated, sluggishly reactive, AC cells and flare present, aphakia	cornea clear, pupil round, regular, reactive, AC quiet, aphakia	cornea clear, pupil round, regular, reactive, AC quiet, aphakia	cornea clear, pupil round, regular, reactive, AC quiet, aphakia	cornea clear, pupil round, regular, reactive, AC quiet, aphakia	17	16.5	16.5	16
	LE	cornea hazy, AC cells present, pupil not reactive, dilated, aphakia	cornea hazy, AC quiet, pupil round, regular, reactive, aphakia	cornea clear, AC quiet, pupil round, regular, reactive, aphakia	cornea clear, AC quiet, pupil round, regular, reactive, aphakia	cornea clear, AC quiet, pupil round, regular, reactive, aphakia	17.5	17	16	15
B	RE	cornea clear, PCIOL in situ, pupil round, regular, reactive, AC quiet	cornea clear, PCIOL in situ, pupil round, regular, reactive, AC quiet	cornea clear, PCIOL in situ, pupil round, regular, reactive, AC quiet	cornea clear, PCIOL in situ, pupil round, regular, reactive, AC quiet	cornea clear, PCIOL in situ, pupil round, regular, reactive, AC quiet	16	17	16.5	16
	LE	cornea hazy, ACIOL in situ, pupil dilated, not reactive, AC cells and flare present, air bubbles present	cornea hazy, ACIOL in situ, pupil round, regular, reactive, AC cells few, air bubbles decreased	cornea clear, ACIOL in situ, pupil round, regular, reactive, AC quiet	cornea clear, ACIOL in situ, pupil round, regular, reactive, AC quiet	cornea clear, ACIOL in situ, pupil round, regular, reactive, AC quiet	17	16.5	16.5	16.5
C	RE	cornea hazy, AC cells present, pupil non-reactive, aphakia	cornea clear, AC quiet, pupil round, regular, reactive, aphakia	cornea clear, AC quiet, pupil round, regular, reactive, aphakia	cornea clear, AC quiet, pupil round, regular, reactive, aphakia	cornea clear, AC quiet, pupil round, regular, reactive, aphakia	17.5	17	18	16.5

	LE	cornea clear, pupil round, regular, reactive, lens normal, AC quiet	cornea clear, pupil round, regular, reactive, lens normal, AC quiet	cornea clear, pupil round, regular, reactive, lens normal, AC quiet	cornea clear, pupil round, regular, reactive, lens normal, AC quiet	cornea clear, pupil round, regular, reactive, lens normal, AC quiet	17	17	17.5	16
D	RE	cornea hazy, AC cells and flare present, SFIOL in situ, pupil non-reactive, dilated	cornea clear, AC cells present, SFIOL in situ, pupil sluggishly reactive	cornea clear, AC cells present, SFIOL in situ, pupil sluggishly reactive	cornea clear, AC cells present, SFIOL in situ, pupil sluggishly reactive	cornea clear, AC cells present, SFIOL in situ, pupil sluggishly reactive	16.5	17	16	16
	LE	cornea clear, pupil sluggishly reactive, cataractous inferiorly dislocated lens, AC quiet	cornea clear, pupil sluggishly reactive, cataractous inferiorly dislocated lens, AC quiet	cornea clear, pupil sluggishly reactive, cataractous inferiorly dislocated lens, AC quiet	cornea clear, pupil sluggishly reactive, cataractous inferiorly dislocated lens, AC quiet	cornea clear, pupil sluggishly reactive, cataractous inferiorly dislocated lens, AC quiet	16	17.5	16.5	16.5

Table 5b. Postoperative Outcomes

‖ Right eye – RE, Left eye – LE, Anterior chamber – AC, Posterior chamber – PC, Intraocular Lens - IOL

DISCUSSION

This prospective study have shown that ectopia lentis were caused by varying predisposing causes among varying age and gender distribution – homocystinuria, ocular trauma and idiopathic. In this study, the cases had presented with wide ranging clinical signs and symptoms, and multiform systemic and ophthalmological findings. The multidisciplinary investigative findings also corroborated the clinical presentations of the cases. The surgical and medical treatment imparted for each case were highly effective, with early and almost complete recovery, and high cure rates.[15] The post-operative prognoses of all the cases had shown favourable outcomes, with almost insignificant complications and adverse effects. Lens abnormalities, mostly ectopia lentis, are an important cause of childhood and adult blindness.[7] Homocystinuria due to remethylation defects is characterised by chronic and progressive neurologic impairment. Homocystinuria due to cystathionine ß-synthase deficiency(chromosome 21q22 mutation), has autosomal recessive inheritance, and manifests with skeletal involvement and ectopia lentis - unlike homocystinuria due to remethylation defects, where plasma methionine concentration is low, and there is neither skeletal involvement nor ectopia lentis.[16] Due to the rare nature of homocystinuria, an inborn error of amino acid metabolism, as well as the associated high morbidity and mortality associated with the untreated natural history, there are very few studies on homocystinuria; this validates the necessity of this study. Along with the corrective refractive surgeries, homocystinuria often responds to a low methionine diet and and adding cysteine, along with high doses of pyridoxine (Vitamin B6) and folic acid.[17,18,19] For recognition, diagnosis and management of homocystinuria due to cystathionine ß-synthase deficiency, a guideline was developed as a part of the European network and registry for homocystinuria and methylation defects (E-HOD), which was further revised, and systematic literature review and evidence grading was done using the Scottish Intercollegiate Guideline Network (SIGN) methodology, resembling the characterisation of Marfan's syndrome by the revised Ghent nosology.[20,21] Systemic manifestations include mental retardation, hypopigmentation of skin and hair, thromboembolic events and marfanoid habitus. Ocular manifestations include myopia and ectopia lentis.[17,18,19] Traumatic ectopia lentis is caused by backward thrust and rebounding lens following blunt trauma to the eye which leads to backward involvement of the iris root by the pressure wave of the aqueous which is usually associated with forcible recoil of the vitreous body. Idiopathic ectopia lentis occurs due to mechanical stretching in cases of buphthalmos, high myopia and so on, or due to inflammatory conditions like zonular destruction and hypermature cataract.

CONCLUSION

Early diagnosis and treatment are essential to prevent visual function. The clinical manifestations and their severity in individual patients may vary considerably and, therefore, the treatment strategy needs to be tailored accordingly. With the advent and development of current adjunctive surgical devices, most of the cases of Ectopia Lentis can now be treated with excellent visual and anatomic results, much

better than what could be accomplished just a few decades ago.

ACKNOWLEDGEMENTS

Sincere acknowledgements to the Department of Ophthalmology, Rajarajeswari Medical College and Hospital, Bangalore, Karnataka, India and Department of Pharmacology, Shri Ramkrishna Institute of Medical Sciences and Sanaka Hospitals, Durgapur, West Bengal, India for the successful completion of this research work and the subsequent compilation of the study literature.

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