Retrospective review of visual outcome in operated lens subluxation

Sultan A. Alzuhairy, MD, Thomas M. Bosley, MD, Abdullah G. Alotaibi, MD.

ABSTRACT

الأهداف: وصف النتائج البصرية للمرضى المصابين بخلع جزئي بعدسة العين الطبيعية يشمل انتباذ العدسة(ectopia lentis) بسبب أمراض جينية والتي تم علاجها جراحياً بالطرق المثبتة والمتعارف عليها عالمياً لكي نتمكن من تقييم فاعلية علاج هذا المرض.

الطريقة: أجريت دراسة استعادية على 17 مريضاً تم تقييمهم وتشخصيهم خضعوا لجراحات تصحيح الخلع الجزئي للعدسة من عام 2000م إلى 2012م بمستشفى الملك عبدالعزيز الجامعي بمدينة الرياض، المملكة العربية السعودية. بسبب ضعف حدة الأبصار الغير ممكن علاجها إلا بالعملية الجراحية. كما أجريت اختبارات المقارنة باختبار الإحصائي.

النتائج: تم إدراج 28 عين بهذه الدراسة وقد شملت المسببات لهذا المرض كالتالي: متلازمة مارفان (12 عين)، وانتباذ العدسة الوراثي (5 أعين)، وبيلة هوموسيستينية (4 أعين)، والأنيميا المنجلية (عينيين الإصابات (6 أعين). تحسن متوسط حدة الإبصار بالإجمال من 20/200 قبل العملية إلى 20/70 بعد العملية($p \geq 0.01$) . وصلت حدة الإبصار ما بعد العملية إلى 20/60≤ أو أحسن في الحالات التي لا يصاحبها كسل بالعين او انفصال بالشبكية أو أي أمراض أخرى موجودة بالعين غير المشكلة الأساسية. كانت نتائج الإبصار بين أنواع العملية الجراحية سواء الحوفي (limbal) أو عن طريق الجسم السطحي (pars plana) كانت متشابهة($p \geq 0.29$) كما أن المرضى المصابين بانتباذ العدسة الوراثي يحصلون على نتائج أفضل في حدة الإبصار من غيرهم ($p \geq 0.01$) ومن ضمن هؤلاء مرضى متلازمة مارفان يحصلون على النتائج الأفضل.

خاتمة: طورت الضوابط والطرق الحديثة المقبولة والمتعارف عليها نجاح عمليات العدسة المخلوعة جزئياً. كما أن المرضى المصابون بخلع جزئي بالعدسة بسبب متلازمة مارفان وبسبب الأمراض الجينية الأخرى يحصلون على النتائج الأفضل بشرط خلو العين من الأمراض الأخرى.

Objectives: To describe the visual outcome of patients with lens subluxation (LS), including ectopia lentis (EL) due to genetic causes, who underwent surgical correction using standard selection criteria and surgical techniques in order to assess effectiveness of current LS therapy.

Methods: This is a retrospective review of 17 sequential patients with LS who underwent lens aspiration between 2000 and 2012 through an anterior (limbal) or posterior (pars plana) approach at King Abdulaziz University Hospital, Riyadh, Saudi Arabia. Snellen visual acuity was converted to the logarithm of the Minimum Angle of Resolution (logMAR) equivalent for statistical analysis. All statistical comparisons were performed by t-test.

Results: This series consisted of 28 eyes with causes of LS including Marfan syndrome (12 eyes), familial EL (5 eyes), homocystinuria (4 eyes), sickle cell anemia (2 eyes), and trauma (6 eyes). Mean visual acuity (VA) for the entire group increased from 20/200 before surgery to 20/70 after surgery ($p \geq 0.01$). Post-operative VA was ≥20/60 in all eyes that did not have complicating factors such as amblyopia, retinal detachment, and/or other ocular abnormalities. Visual outcome of limbal and pars plana approaches was statistically similar ($p \geq 0.29$). Patients with genetic causes of EL had a significantly better visual outcome than other patients ($p \geq 0.01$); out of these, patients with Marfan syndrome had a better visual outcome than other patients with genetic abnormalities ($p \geq 0.02$).

Conclusions: Accepted surgical criteria and techniques improved visual outcome in patients with LS. Patients with Marfan syndrome and others genetic abnormalities without ocular complications had best visual outcomes.

Saudi Med J 2013; Vol. 34 (10): 1030-1034

From the Ophthalmology Department, College of Medicine, King Saud University, Riyadh, Kingdom of Saudi Arabia.

Received 18th June 2013. Accepted 19th August 2013.

Address correspondence and reprint request to: Dr. Sultan A. Alzuhairy, Department of Ophthalmology, College of Medicine, Qassim University, PO Box 4490, Buraidah 51491, Kingdom of Saudi Arabia. Tel. +966 555134777. Fax. +966 (11) 4775724. E-mail: dr.sulzuh@gmail.com

Lens subluxation (LS) is displacement of the lens from its normal position that may be acquired because of trauma or may be congenital or developmental ectopia lentis (EL).[1] Non-traumatic EL is associated with Marfan syndrome,[2] homocystinuria, aniridia, congenital glaucoma, and less frequently with Ehlers-Danlos syndrome, hyperlysinemia, and sulfite oxidase deficiency. It can also occur as an isolated familial autosomal dominant trait.[1] Lens subluxation can cause decreased vision, marked astigmatism, monocular diplopia, and iridodonesis.[1] Other potential complications include cataract; pupillary block and angle-closure glaucoma from displacement of the lens into the anterior chamber; or subluxation of the lens posteriorly into the vitreous cavity, which often has no adverse sequela.[1] Historically, surgical management of LS was avoided because of associated intraoperative and postoperative complications.[3] The development of limbal[3] and pars plana[4,5] microsurgical techniques since the 1970s has permitted the surgical treatment of LS with lower complication rates and better outcomes.[6] We evaluated visual outcome in a sequential group of patients selected for LS surgery using current standard criteria in order to compare visual acuity before and after surgery and to compare visual outcome between patients with traumatic and non-traumatic causes of LS.

Methods. King Saud University College of Medicine Institutional Review Board approved this retrospective review of 17 sequential patients with LS who underwent lens aspiration between 2000 and 2012 through an anterior (limbal) or posterior (pars plana) approach at King Abdulaziz University Hospital in Riyadh, Saudi Arabia. This research followed the principles of the Helsinki Declaration. All patients were followed pre-operatively or post-operatively by one pediatric ophthalmologist (AGA). Only one operated patient was excluded because the patient failed to return for post-operative examination.

Chart information was collected regarding personal and family history; the results of a comprehensive pre-operative ophthalmologic examination including best corrected Snellen visual acuity (VA), refraction, measurement of intraocular pressure (IOP), slit lamp examination of the anterior segment, and a dilated fundus exam; post-operative examination including VA, IOP, slit lamp examination, manifest and cycloplegic refraction, and fundus examination; and the results of a pediatric consultation for signs of syndromes associated with EL. Diagnosis of associated genetic problems was made by general medical criteria.

Indications for lens aspiration in patients with LS included VA less than 20/60 with glasses, monocular diplopia, lens displacement into either the anterior or posterior chamber, lens-induced uveitis or glaucoma, and impending subluxation of the lens.[4,7] Selection of surgical approach depended on lens status. A pediatric ophthalmologist performed lens aspiration or extracapsular cataract extraction (ECCE) with anterior vitrectomy with or without IOL implantation through an anterior approach if the lens was accessible,[4] while a retinal surgeon used a pars plana approach with pars plana vitrectomy if an anterior approach was not possible.[3] All patients under 10 years old had an anterior vitrectomy when operated from the anterior approach. Visual rehabilitation consisted of intraoperative intraocular lens (IOL) implantation or postoperative glasses or contact lens.

Snellen visual acuity was converted to the logarithm of the Minimum Angle of Resolution (logMAR) equivalent for statistical analysis. All statistical comparisons were performed by t-test (SPSS version 19, International Business Machines Corp, Armonk NY USA). The diagnosis of amblyopia was made in an amblyogenic circumstance when optotype acuity was reduced by 2 lines or more when compared to the contralateral eye or to 20/60 or worse in patients with bilateral disease.[8] Patients with amblyopia were treated with glasses or patching both before and after surgery.

Results. **Table 1** contains demographic and basic ophthalmologic information regarding each of the 28 eyes operated for LS. Duration of follow-up ranged from 6-120 months (mean 42 months). Bilateral surgery was performed in 11 patients and unilateral surgery was performed in 6. Post-operative optical correction consisted of glasses in 23 eyes (82%), intraocular lens in 4 eyes (14%), and contact lens in one eye (4%). Pre-operatively, 4 eyes had amblyopia, 2 eyes had retinal detachment, and 3 eyes had other ocular complications. Postoperative improvement in VA to ≥20/60 occurred in 17 eyes (61%), including every eye without the above complications. Visual acuity deteriorated in only one eye, which had a pre-operative retinal detachment (4%). Final VA (mean 20/70) was significantly better than initial VA (mean 20/200) for all operated eyes considered together ($p \geq 0.01$). Fifteen eyes underwent pars plana lensectomy and vitrectomy because of associated retinal

Disclosure. Authors have no conflict of interests, and the work was not supported or funded by any drug company.

detachment in 3 eyes, lens subluxation into the vitreous in 5 eyes, and severe lens subluxation in 7 eyes. These eyes had a post-operative VA that was not statistically different from patients operated through a limbal approach ($p \geq 0.29$). Patients with inherited causes of EL (Marfan syndrome, homocystinuria, SSD, and familial EL) had significantly better initial ($p \geq 0.01$) and final ($p \geq 0.01$) VA than patients with LS due to trauma. Patients with Marfan syndrome had statistically similar initial VA as other patients with inherited causes of EL ($p \geq 0.11$), but their post-operative VA was significantly better ($p \geq 0.02$).

Discussion. We describe the outcome of surgery for LS on 28 sequential eyes of 17 patients cared for at a national ophthalmological center over a 12 year period. Patients were evaluated and operated only if an adequate visual outcome could not be achieved by conservative visual rehabilitation. All received rapid pre-operative evaluation, which was important for better visual outcome because some children may develop amblyopia. A thorough follow-up ophthalmologic assessment with refraction was also fundamental.[9] Prompt referral to a pediatrician or general practitioner was obtained because the ophthalmologist was often first physician to become aware of a patient with EL due to an associated systemic disease that required independent management.[10] Genetic causes of EL were more common than traumatic causes in this patient group, with the most common systemic diseases

Table 1 - Demographics and basic ophthalmologic information of the 28 eyes operated for lens subluxation (LS).

Patient/eye	Gender, age at presentation	Cause	Initial VA	Final VA	Surgery	Comments
1, OD	F, 11	Marfan	20/100	20/30	PPL + PPV	
1, OS	F, 11	Marfan	20/100	20/20	PPL + PPV	
2, OD	M, 3	Marfan	CF	20/20	lens aspiration + ant vitrectomy + PC IOL	
2, OS	M, 3	Marfan	CF	20/20	lens aspiration + ant vitrectomy + PC IOL	
3, OD	F, 12	Marfan	20/200	20/20	lens aspiration + ant vitrectomy	
3, OS	F, 12	Marfan	20/200	20/20	lens aspiration + ant vitrectomy	
4, OD	F, 8	Marfan	20/100	20/25	PPL + PPV	
4, OS	F, 8	Marfan	20/100	20/30	PPL - PPV	
5, OD	M, 26	Marfan	20/100	20/40	PPL + PPV	IOP high on presentation but normal after surgery
5, OS	M, 26	Marfan	CF 3ft	20/50	PPL + PPV	
6, OD	M, 29	Marfan, trauma	20/160	20/60	PPL + PPV	
6, OS	M, 29	Marfan	20/400	CF 5feet	PPL + PPV + RD repair	pre-operative retinal detachment
7, OD	M, 5	Homocystinuria		F/F	PPL + PPV	uncooperative
7, OS	M, 5	Homocystinuria		F/F	PPL + PPV	uncooperative
8, OD	M, 8	Homocystinuria		20/30	lensectomy + ant vitrectomy	
8, OS	M, 8	Homocystinuria		20/40	PPL+PPV	
9, OD	M, 11	SSD	20/40	20/40	lens aspiration + ant vitrectomy	
9, OS	M, 11	SSD	20/40	20/40	lens aspiration + ant vitrectomy	
10, OS	M, 5	Trauma	HM	20/200	lens aspiration + ant vitrectomy	amblyopia, non-compliant with contact lens
11, OS	M, 21	Trauma	HM	CF	ECCE + ant vitrectomy	fibrous tissue covering macula
12, OD	M, 13	Trauma	CF	CF	PPL + PPV with retinal reattachment surgery with silicon oil	rhegmatogenous retinal detachment before surgery
13, OS	M, 3	Trauma		NLP	PPL + PPV	lost to follow up for 12 years after secondary repair
14, OS	F, 3	Trauma	CF	20/100	lens aspiration + ant vitrectomy + CTR + PC IOL	corneal laceration and delayed management leading to amblyopia
15, OD	M, 20	Familial	20/80	20/40	lens aspiration + ant vitrectomy	
15, OS	M, 20	Familial	20/80	20/60	lens aspiration + ant vitrectomy	
16, OS	F, 3	Familial	CF	CF	lens aspiration + ant vitrectomy + AC IOL	amblyopia
17, OD	F, 3	Familial		20/80	PPL + PPV	
17, OS	F, 3	Familial		CF	PPL + PPV	amblyopia

OD - right, OS - left, M - male, F - female, SSD - sickle cell disease, HM - hand motions, CF - counting fingers, PPL - pars plana lensectomy, PPV - pars plana vitrectomy, ant - anterior, CTR - capsular tension ring, ECCE - extracapsular cataract extraction, VA - visual acuity, IOL - intraoperative intraocular lens, PC - posterior chamber, HM - hand motion, AC - anterior chamber

being Marfan syndrome (12 eyes) and homocystinuria (4 eyes).

Benefits and risks of surgery were assessed prior to surgery because some patients had associated ocular problems such as retinal detachment and amblyopia that predicted a poor visual outcome. Final VA for the group as a whole was significantly better than initial VA, and every patient without additional ocular complications had an excellent visual outcome after surgery and visual rehabilitation. Both limbal and pars plana approaches were employed depending on the status of the lens and accessibility of the lens through a limbal incision, and visual outcome of these 2 approaches was statistically similar. Patients with genetic causes of EL did better than those with trauma, although visual assessment of patients with homocystinuria was limited by the patients' cognitive function. Traumatized eyes had a worse visual prognosis than non-traumatized eyes, especially if the trauma was associated with other ocular problems such as retinal detachment or corneal laceration. These results are a testimony to the effectiveness of surgical advances over the last several decades.

Final visual outcome was heavily dependent on visual rehabilitation. Axial myopia is frequent in EL,[11] and methods for visual rehabilitation postoperatively include glasses, contact lenses, and intraocular lenses. Shortt et al[4] described improved vision in 22 out of 24 eyes with postoperative glasses or contact lens, and Anteby et al[3] concurred that correction of aphakia with glasses or contact lenses was safer than scleral fixated or anterior chamber intraocular lens. However, children are not always compliant with wearing glasses, and contact lenses demand maintenance and can result in complications such as keratitis, corneal vascularization, or corneal perforation. In the current study optical lens correction with glasses was used most commonly, followed by IOL implantation, and then contact lens.[6] The study population is too small for statistical comparison of the visual outcome of different types of optical correction.

Additional surgical approaches are sometimes necessary in patients with EL. Several techniques have been introduced to offer good IOL centration to the patient with zonular weakness. Konradsen et al reported good postoperative visual outcomes with capsular tension ring (CTR),[7] and several other authors have reported satisfactory results with IOL implantation with or without CTR.[12] In the current study, CTR was used in one patient with satisfactory improvement of visual outcome. Other studies have urged caution with anterior chamber IOLs in pediatric patients due to the increased risk over time of corneal endothelial damage and angle-closure glaucoma,[3] and anterior chamber IOL was used in only one patient in this series. Scleral sutured fixation of posterior chamber IOL is a safe technique, but postoperative complications after surgery are not uncommon.[3] This approach was not necessary in our patient series.

The best visual outcome in this patient series was in patients with Marfan syndrome. Ectopia lentis occasionally may be the only sign of Marfan syndrome,[10] but usually other aspects of the phenotype make a clinical diagnosis practical.[2] As is commonly the case,[1] none of the patients described here were known to have Marfan syndrome prior to an ophthalmologic diagnosis of EL. This diagnosis is important because the risk of retinal detachment associated with EL in Marfan syndrome has been reported to be between 10 and 25%; in fact, one of 12 eyes in Marfan patients in the current group had both a retinal detachment and a sub-luxated lens. Visual outcome of EL surgery in Marfan syndrome is generally good if the patient presents early with no associated ocular abnormalities, but some potential complications are unique to this disorder.[13]

In conclusion, approaches to visual rehabilitation of patients with LS have changed substantially over recent decades, particularly because of newer microsurgical techniques. Many patients can be managed with only optical correction; however, surgery is necessary at times because the visual outcome of conservative management is not adequate. This relatively small retrospective series from one ethnic group shows that applying current criteria for surgery together with postoperative visual rehabilitation resulted in a significant improvement in visual outcome with best improvement being reserved for patients without associated ocular abnormalities. Early evaluation, rigorous visual rehabilitation, and management of general medical problems associated with certain genetic etiologies are all important to the ultimate outcome.

References

1. Merin S. Inherited Eye Disease. New York (NY): Marcel Dekker; 1991.
2. Faivre L, Masurel-Paulet A, Collod-Beroud G, Callewaert BL, Child AH, Stheneur C. Clinical and molecular study of 320 children with Marfan syndrome and related type I fibrillinopathies in a series of 1009 probands with pathogenic FBN1 mutations. ***Pediatrics*** 2009; 123: 391-398.

3. Anteby I, Isaac M, BenEzra D. Hereditary subluxated lenses: visual performances and long-term follow-up after surgery. ***Ophthalmology*** 2003; 110: 1344-1348.
4. Shortt AJ, Lanigan B, O'Keefe M. Pars plana lensectomy for the management of ectopia lentis in children. ***J Pediatr Ophthalmol Strabismus*** 2004; 41: 289-294.
5. Wu-Chen WY, Letson RD, Summers CG. Functional and structural outcomes following lensectomy for ectopia lentis. ***J AAPOS*** 2005; 9: 353-357.
6. Kim SY, Choung HK, Kim SJ, Yu YS. Long-term results of lensectomy in children with ectopia lentis. J Pediatr Ophthalmol Strabismus 2008; 45: 13-19.
7. Konradsen T, Kugelberg M, Zetterström C. Visual outcomes and complications in surgery for ectopia lentis in children. ***J Cataract Refract Surg*** 2007; 33: 819-824.
8. Panel APOS. Amblyopia. San Francisco (CA): American Academy of Ophthalmology; 2007.
9. Park SC, Chung ES, Chung TY, Kim SA, Oh SY. Axial growth and binocular function following bilateral lensectomy and scleral fixation of an intraocular lens in nontraumatic ectopia lentis. ***Jpn J Ophthalmol*** 2010; 54: 232-238.
10. Zadeh N, Bernstein JA, Niemi AK, Dugan S, Kwan A, Liang D, et al. Ectopia lentis as the presenting and primary feature in Marfan syndrome. ***Am J Med Genet A*** 2011; 155A: 2661-2668.
11. Romano PE, Kerr NC, Hope GM. Bilateral ametropic functional amblyopia in genetic ectopia lentis: its relation to the amount of subluxation, an indicator for early surgical management. ***Binocul Vis Strabismus Q*** 2002; 17: 235-241.
12. Ozdek S, Sari A, Bilgihan K, Akata F, Hasanreisoglu B. Surgical treatment of hereditary lens subluxations. ***Ophthalmic Surg Lasers*** 2002; 33: 309-313.
13. Vasavada AR, Praveen MR, Desai C. Management of bilateral anterior dislocation of a lens in a child with Marfan's syndrome. ***J Cataract Refract Surg*** 2003; 29: 609-613.

Related Articles

Al-Khatib TK, Al-Khulaidi NA. Traumatic subconjunctival crystalline lens. Dislocation by the tail of a cow. ***Saudi Med J*** 2004; 25: 2018-2019.

Osman EA, Al-Fawaz N, Al-Otaibi AG, Al-Mansouri SM, Mousa A, Al-Mezaine HS. Glaucoma after open globe injury at a tertiary care university hospital in Central Saudi Arabia. Cumulative incidence and risk factors. ***Saudi Med J*** 2013; 34: 374-378.

Akar S, Dogan M, Kucuksumer Y, Kaya V, Bayraktar S, Gokyigit B, et al. Long-term results of phakic anterior chamber intraocular lens implantation in myopic eyes. ***Saudi Med J*** 2010; 31: 939-941.