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Lens-sparing vitrectomy for shaken baby syndrome

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保留晶状体的玻璃体切除术治疗婴儿摇荡综合征

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摘要

目的:分析婴儿摇荡综合征(SBS)患者黄斑前及玻璃体出血遮挡黄斑的眼部特征及视力。患者均接受保留晶状体的玻璃体切除术(LSV)。

方法:回顾性研究。选取2010年至2012年确诊为黄斑前及玻璃体出血遮挡黄斑的SBS并接受LSV的患者,对其眼部特征和人口统计学数据进行分析。均数的比较采用配对t检验,分析分类数据采用Fisher精确检验和Pearson卡方检验。$P<0.05$有统计学意义。

结果:共32例患者纳入本研究,平均年龄为5.09±1.96mo。在本研究中,21例(65.6%)患者视力为无光感。患者瞳孔较大者初始视力较差($P=0.021$),且大部分接受过神经外科手术($P=0.027$)。行LSV的平均间隔为28.56±20.83d。我院玻璃体切除术的并发症发生率为4.26%。术后26例(80%)患者视力为光感或更好,术后等效球镜大多为近视($P=0.001$)。

结论:SBS患者的眼科评估对于最佳视觉预后非常重要。对于未清除眼内出血致黄斑模糊的SBS患者,LSV的并发症发生率较低,不失为早期手术治疗中的好选择。

关键词:玻璃体切除术;婴儿摇荡综合征;黄斑出血;玻璃体出血

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Abstract

• AIM: To document the ophthalmological findings and visual outcomes for shaken baby syndrome (SBS) patients who had lens-sparing vitrectomy (LSV) for non-resolving premacular and vitreous hemorrhages obscuring the macula.

• METHODS: A retrospective review and statistical analysis of ophthalmological and demographic data of patients admitted with SBS from 2010 to 2012 was done. Patients with premacular and vitreous hemorrhage obscuring the macula who underwent LSV were included as subjects. Paired samples t-test was used to compare means, categorical data was analysed using Fisher's exact test and Pearson Chi-squared test. P value of less than 0.05 was considered as statistically significant.

• RESULTS: Thirty two subjects were recruited with a mean age of 5.09±1.96mo. Twenty one (65.6%) subjects had visual acuity of no light perception at presentation. Subjects with poorer initial visual acuity had larger pupil sizes ($P=0.021$) and most of them had neurosurgical intervention ($P=0.027$). The mean duration to perform LSV was 28.56±20.83d. Our vitrectomy complication rate was 4.26%. Post-operatively, 26 (80%) subjects had vision of light perception or better, the spherical equivalent was significantly more myopic ($P=0.001$).

• CONCLUSION: Prompt ophthalmological assessment is vital to ensure optimum visual rehabilitation in SBS patients. With low complication rates, early surgical intervention with LSV represents a promising option for non-resolving intraocular hemorrhages obscuring the macula in SBS.

• KEYWORDS: vitrectomy; shaken baby syndrome; macula hemorrhage; vitreous hemorrhage
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INTRODUCTION

Shaken baby syndrome (SBS) causes a significant amount of morbidity and mortality to the paediatric population. SBS is diagnosed by the triad of retinal hemorrhages, subdural hemorrhage and encephalopathy[1]. It is reported that 64-70% of SBS patients never fully recover from their injuries sustained and 15-38% of SBS patients die during their hospital admission[2-3]. Abnormal vision has been reported in 65% of SBS survivors[4]. Premacular and vitreous hemorrhages in SBS frequently obscure the macula and become an amblyogenic threat to the developing eye. Lens-sparing vitrectomy (LSV) has been suggested as a viable option to treat persistent visually-depriving hemorrhages in children as it reduces the rate of cataract formation and negates the need for lensectomy for surgical access to the pars pliacata region[5-7]. Limited data however exists regarding the eventual visual outcomes and safety profile of LSV in SBS. This study aims to document the ophthalmological findings and visual outcomes for SBS patients who had LSV for non-resolving premacular and vitreous hemorrhages obscuring the macula.

SUBJECTS AND METHODS

This retrospective study was conducted in Hospital Kuala Lumpur (HKL) from Nov. 1st 2012 to Mar. 1st 2013. The study and data collection conformed to all local laws and was compliant with the guidelines of the Declaration of Helsinki. Ethical approval was obtained from the Medical Research and Ethics Committee of the Malaysian Ministry of Health (NMRR ID NMRR-13-359-15951 S1 R0). All consecutive patients admitted to the pediatric and neurosurgical wards with a diagnosis of subdural haemorrhage secondary to suspected SBS from Jan. 1st 2010 to Dec. 31st 2012 were identified from ward registers. The patients were recruited as study subjects if they had documented evidence of premacular or vitreous hemorrhage confirmed by ophthalmological assessment and had LSV. The patients were excluded if they had trauma or disease which caused the intraocular or subdural hemorrhage. The patients were also excluded if they passed away during admission or did not have a minimum of 1mo ophthalmological follow-up postoperatively.

The criterion for subjects to undergo LSV was premacular or vitreous hemorrhage obscuring the macula which did not spontaneously resolve within a period of 4wk of observation or severe vitreous hemorrhage which completely obscured fundal view. All the LSV surgeries were performed under general anaesthesia by a single surgeon. The distance of the sclera incision from the corneal limbus ranged from 1.5 mm to 2.0 mm depending on the age of the child. A three-port, trans pars plana vitrectomy (23 gauge system) using Topcon Offis Wide angle viewing system was used according to the method already described in previous literature[6].

Epidemiological data consisted of the patients' ages, gender, ethnic group, primary caregiver, birth history, medical problems, family members, presenting symptoms, medical and neurosurgical treatment and eventual outcomes. Ophthalmological data collected included presenting visual acuity, documented eye findings, ocular surgical intervention and final visual acuity after a minimum of 3mo follow-up. For all subjects, data from one eye was taken for data analysis. The eye chosen was the eye which underwent LSV. If both eyes had LSV, then the worst eye was chosen for analysis. Visual acuity at presentation was divided into visual acuity of perception of light or non-perception of light. Pupil measurements were taken using a ruler. Anterior segment examination was done using a portable slit lamp and binocular indirect ophthalmoscopy was utilised for fundus examination. Intraocular pressure was measured using tonopen. All data was entered into a standardized data collection sheet and analyzed by an ophthalmologist. No new investigations were ordered and all data was handled anonymously. There was no communication between the researcher and the patient, the patient's family, the patient's treating physicians or the investigating police officers or social welfare officers during the course of data collection.

The data collected was entered into an Excel spreadsheet. SPSS version 13 (SPSS Statistics: Windows Student Version 13, Chicago, Illinois, USA) was used for data analysis. Paired samples t-test was used to compare means and categorical data was analysed using Fisher's exact test and Pearson Chi-squared test. P value of less than 0.05 was considered as statistically significant.

RESULTS

The number of patients admitted with suspected SBS was 17, 23 and 36 patients in the year 2010, 2011 and 2012 respectively. A total of 32 subjects were recruited from the 76 patients admitted with suspected SBS and data from 32 eyes were used for statistical analysis. Nine patients were excluded as they had passed away during admission and 35 patients were excluded as they did not require LSV. The subjects recruited represented 42.11% of all patients admitted to HKL for suspected SBS within a 3-year period.

The subjects' ages ranged from 2 to 11mo of age with a mean age of 5.09mo (SD: 1.96). More than half of the subjects (59.4%) were males. A majority of the subjects (90.6%) were Malay, followed by Chinese patients. Only 1 subject was born prematurely. The mean birth weight of the subjects was 2.97 kg (SD: 0.49). The medical history of all subjects was unremarkable except for 1 subject who had Down's syndrome. Almost half of the subjects (47.2%) did not have siblings while the remaining subjects that had siblings were the youngest

Table 1 Pre-operative visual acuity, pupillary size and subjects requiring neurosurgical intervention

Pre-operative details	Visual acuity		*P*
	Better than NPL	NPL	
Pupil size ($n=32$)			
<4mm	8	5	0.021
≥4mm	3	16	
Neurosurgical intervention ($n=32$)			
Yes	3	15	0.027
No	8	6	

PL: Perception of light; NPL: Non-perception of light.

in their respective families. Most of the subjects (81.8%) were in nursery or in the care of a babysitter when the incident occurred.

Fifty percent (16/32) of the subjects presented with seizures and 37.5% (12/32) of subjects were found unresponsive at the time of presentation to hospital. The majority of the subjects had a Glasgow Coma Scale (GCS) score of less than 8 at the time of hospital admission. Ninety point six percent (29/32) subjects had their first ophthalmological assessment within 10d of hospital admission. The pupillary size of the subjects ranged from 2 mm to 8 mm with a mean of 4.91 (SD: 1.86). No documentation was available with regards to environment illumination during pupillary size measurement. The subjects presenting spherical equivalent was plano to +5.50 D with a mean of 1.68 D (SD: 1.54).

All subjects had normal anterior segment findings and normal intraocular pressure. Twenty-nine subjects (90.6%) had bilateral intraocular haemorrhages. Out of the 61 eyes with intraocular hemorrhage, 22 eyes were noted to have traumatic macular retinoschisis from fundus examination. Ten eyes presented with vitreous hemorrhage. Upon first presentation, 65.6% (21/32) subjects had visual acuity of non-perception of light (NPL) while 34.4% (11/32) subjects had visual acuity of perception of light. Our study found that subjects with presenting visual acuity of NPL had larger pupils ($P=0.021$) and more of them had neurosurgical intervention ($P=0.027$) in comparison to subjects with visual acuity of better than NPL. (Table 1)

The duration to perform LSV ranged from 8 to 90d post ocular diagnosis with a mean of 28.56d (SD: 20.83). Seventy-eight point one percent (25/32) of subjects had LSV within 30d post ocular diagnosis. Four subjects had delayed surgery beyond 2mo as 2 subjects were intially unfit for surgery and 2 other subjects had been treated at other health facilities before being transferred to our center for non-resolving vitreous hemorrhage. Out of the 61 eyes presenting with intraocular hemorrhage, 47 eyes underwent LSV. Two of the 47 eyes that underwent vitrectomy had surgical complications. One eye developed retinal detachment post-operatively which was treated with circumferential scleral buckling and laser. The subject's eye later developed cataract and plain lens aspiration was done in view of the poor visual prognosis. Another subject's eye had an iatrogenic retinal break during peeling of the internal limiting membrane. The retinal break was treated successfully with barricade laser and the subject had good post-operative vision of 6/24 at 3y follow-up. Twenty-one point eight percent (7/32) subjects underwent simultaneous bilateral LSV. No cases of endophthalmitis or persistently raised intraocular pressure were reported.

Table 2 Final visual outcomes of lens-sparing vitrectomy

Final Visual outcomes	Preop	Postop	*P*
Visual acuity			
NPL	21 (65.6%)	6 (18.75%)	
PL or better	11 (34.4%)	26 (81.25%)	0.000
SE($\bar{x}\pm s$)	1.68±1.54	−1.03±1.91	0.001

NPL: No perception of light; PL: Perception of light; SE: Spherical equivalent.

The follow-up of subjects post-operatively ranged from 1 to 48mo (Mean: 10.75, SD: 10.97). Eighty percent of the subjects (26/32) had vision of perception of light or better, and the post-operative spherical equivalent was significantly more myopic in comparison to the spherical equivalent at presentation (Table 2). Fifty-six point three percent (18/32) subjects required neurosurgical intervention for intracerebral hemorrhage and 78.1% (25/32) of subjects had signs of developmental delay.

DISCUSSION

An increasing number of patients with suspected SBS were admitted to our centre every year. This trend is worrying as SBS is a preventable tragedy. The increase may be an actual increase or secondary to heightened awareness regarding SBS. Urbanisation of Malaysia has resulted in more women entering the workforce. This has caused an increase of childcare by babysitters or nurseries, with many having untrained caregivers who are ill prepared to handle young infants[8]. Our study noted that babysitters and nurseries made up more than

80% of the primary caregivers of the affected children. Our SBS subjects' ages ranged from 2 to 11mo and majority of the subjects were male patients. This corresponded to published reports on SBS[4,9]. Prematurity was not a risk factor for SBS as the majority of our study subjects were born at full term. This contrasted with studies which cited prematurity as a risk factor for SBS[10]. In addition, only 1 of the 32 patients was developmentally delayed prior to hospital admission.

Most of our subjects were admitted with seizures, or were found unresponsive at first hospital presentation. This was consistent with published clinical presentations of SBS[11]. Larger pupillary size was reported to be associated with poorer survival in trauma patients[12]. We noted that a pupillary size of 4 mm or larger was associated with poorer visual acuity at initial hospital presentation. This may reflect the increased severity of ocular and cerebral trauma inflicted, and explain why more of these subjects had neurosurgical intervention.

The duration from the time of admission until an ophthalmological assessment was performed ranged from 0 to 15d. Reasons for delayed eye assessment included the unstable condition of patients, misdiagnosis or missed eye referrals during transfer between wards or hospitals. An increased awareness for an examination by an ophthalmologist is essential as missed retinal haemorrhages may lead to missed diagnosis of SBS. In previous literature, non-ophthalmologists were only able to identify 87% of retinal hemorrhages in SBS patients[13].

A majority of subjects were noted to have bilateral intraocular hemorrhages. This was consistent with Maguire and colleagues who noted a predominance of bilateral retinal hemorrhages in SBS[14]. Vitreous hemorrhage is an uncommon but significant finding in SBS as it has the most evidence linking it to SBS. It is believed to be secondary to progressive breakthrough bleeding of retinal hemorrhages into the vitreous over a number of days and is not thought to be an acute event[15]. We noted that SBS subjects presenting with vitreous hemorrhage frequently required LSV as the vitreous hemorrhage was non-resolving. The non-resolution of vitreous hemorrhage in SBS may be due to injury chronicity and poor blood absorption by the infantile vitreous.

Early visual stimulation is crucial for the development of neural infrastructure for visual learning[16]. A minimum vision of perception of light is required for circadian rhythms and maximizing the potential for cognitive skills in children[17]. Hence, any form of visual deprivation regardless of duration will have a lasting impact on a child's visual development even after the deprivation has ended. This emphasizes the necessity for early removal of intraocular haemorrhages obscuring the macula to reduce the amblyogenic potential of these hemorrhages. In addition, early removal of such haemorrhages has a protective effect on the macula as late retinal scarring has been documented in SBS patients due to the migration of blood from the retina to the vitreous and excess iron causes oxidative damage to the retina[18-19]. With the advent of LSV, the treatment of amblyogenic, persistent intraocular haemorrhage is surgical. The ideal time for LSV for SBS is not known but it is suggested that earlier surgical intervention may be best between 11 to 28d post ocular diagnosis in a child with SBS[20]. The majority of our study subjects had LSV within 30d of ocular diagnosis.

We noted that early intervention for non-resolving hemorrhages obscuring the macula allowed for earlier visual rehabilitation. This was supported by the fact that even though 65.6% (21/32) subjects had poorer vision at presentation, 81.25% (26/32) subjects had a visual acuity of perception of light or better post LSV. Twenty out of the 32 cases who went for vitrectomy had normal fundi post-operatively. Although only 5 subjects managed to achieve vision better than 6/24 at 3y follow-up, the final visual acuity may be much better should the subjects be followed up longer. Our vitrectomy complication rate was 4.26% where 2 of the 47 vitrectomised eyes developed complications. This result is better in comparison to the study by Capone[6] who noted a complication rate of 2 out of 16 eyes (12.5%) which underwent LSV for infantile vitreous haemorrhage. Of the 10 subjects that completed a post-operative follow-up of 6mo or longer, there was no documented cataract or development of glaucoma noted.

Myopic progression was noted in our study subjects post LSV. This myopic progression could be similar to the myopic progression seen in adults post vitrectomy, where the myopic shift was secondary to corneal or sclera stretching, anterior chamber depth changes and silicone oil tamponade[20]. Also, the infantile macula has been noted to be more vulnerable to the presence of blood leading to progressive myopia[21]. In addition, the presence of dense vitreous hemorrhage could have caused stretching of the cornea and sclera leading to corneal enlargement and increase in axial length[20]. This could not be confirmed in our study as no A-scans were done to document the axial length pre- and post-vitrectomy.

A majority of the subjects were eventually noted to be developmentally delayed. This may be due to the chronicity and increased severity of the ocular and brain injuries sustained. The chronicity and increased severity of the brain injuries may also explain why some subjects had later eye assessments as the subjects may have been medically unstable for eye examination and may have been transferred to multiple wards leading to missed diagnosis.

The limitations of our study were that it was a retrospective study comprising of a small number of patients. The follow-up

was short and data collection was dependant on the available documentation. Documentation regardig pupillary responses and afferent pupillary defect were sparse due to the difficulty of checking relative afferent pupillary defect in children. Also, investigations like visual evoked potential, electroretinogram and visual fields were not performed for quantitative visual assessment. Despite the limitations, our study is the largest to date and the first in reporting the results of LSV for premacular and vitreous hemorrhage obscuring the macula for SBS patients in Malaysia. HKL is also the sole tertiary referral centre for LSV for children in Malaysia and may accurately reflect the presentation of SBS patients requiring LSV in the urban Malaysian population.

With low complication rates, our study suggests that early surgical intervention with LSV is a viable option for non-clearing premacular and vitreous hemorrhages obscuring the macula in SBS. This study also highlights the need for early ophthalmological assessment to ensure optimum visual rehabilitation in SBS patients.

REFERENCES

1 Squier W. The "Shaken Baby" syndrome: pathology and mechanisms. *Acta Neuropathologica* 2011;122(5):519-542

2 Fanconi M, Lips U. Shaken baby syndrome in Switzerland: results of a prospective follow-up study, 2002-2007. *Eur J Pediatr* 2010;169(8):1023-1028

3 Haviland J, Russell RI. Outcome after severe non-accidental head injury. *Arch Dis Child* 1997;77(6):504-507

4 King WJ, MacKay M, Sirnick A; Canadian Shaken Baby Study Group. Shaken baby syndrome in Canada: clinical characteristics and outcomes of hospital cases. *CMAJ* 2003;168(2):155-159

5 Rahman W, Osbourne S, Bhan A, Orr GM, Richard Gregson R. Vitrectomy in cases of nonaccidental injury. *J Pediatr Ophthalmol Strabismus* 2010;47(3):163-167

6 Capone A. Lens-sparing vitreous surgery for infantile amblyogenic vitreous hemorrhage. *Retina (Philadelphia, Pa)* 2003;23(6):792-795

7 Maguire AM, Trese MT. Visual results of lens-sparing vitreoretinal surgery in infants. *J Pediatr Ophthalmol Strabismus* 1993;30(1):28-32

8 Kasim MS, Cheah I, Shafie HM. Childhood deaths from physical abuse. *Child Abuse Negl* 1995;19(7):847-854

9 Thalayasingam M, Veerakumarasivam A, Kulanthayan S, Khairuddin F, Cheah IG. Clinical clues for head injuries amongst Malaysian infants: accidental or non-accidental? *Injury* 2012;43(12):2083-2087

10 Altimier L. Shaken baby syndrome. *J Perinat Neonatal Nurs* 2008;22(1):68-76

11 Carbaugh SF. Understanding Shaken Baby Syndrome. *Adv Neonatal Care* 2004;4(2):105-114

12 Lieberman JD, Pasquale MD, Garcia R, Cipolle MD, Mark Li P, Wasser TE. Use of admission Glasgow Coma Score, pupil size, and pupil reactivity to determine outcome for trauma patients. *J Trauma* 2003;55(3):437-442

13 Morad Y, Kim YM, Mian M, Huyer D, Capra L, Levin AV. Nonophthalmologist accuracy in diagnosing retinalhemorrhages in the shaken baby syndrome. *J Pediatr* 2003;142(4):431-434

14 Maguire SA, Watts PO, Shaw AD, Holden S, Taylor RH, Watkins WJ, Mann MK, Tempest V, Kemp AM. Retinal haemorrhages and related findings in abusive and non-abusive head trauma: a systematic review. *Eye (Lond)* 2013;27:28-36

15 Togioka BM, Arnold MA, Bathurst MA, Ziegfeld SM, Nabaweesi R, Colombani PM, Chang DC, Abdullah F. Retinal hemorrhages and shaken baby syndrome: an evidence-based review. *J Emerg Med* 2009;37(1):98-106

16 Lewis TL, Maurer D. Effects of early pattern deprivation on visual development. *Optom Vis Sci* 2009;86(6):640-646

17 Walsh MK, Drenser KA, Capone A, Trese MT. Early vitrectomy effective for Norrie disease. *Arch Ophthalmol* 2010;128(4):456-460

18 Greenwald MJ, Weiss A, Oesterle CS, Friendly DS. Traumatic retinoschisis in battered babies. *Ophthalmology* 1986;93(5):618-625

19 He X, Hahn P, Iacovelli J, Wong R, King C, Bhisitkul R, Massaro-Giordano M, Dunaief JL. Iron homeostasis and toxicity in retinal degeneration. *Prog Retin Eye Res* 2007;26(6):649-673

20 Tseng PC, Woung LC, Tseng GL, Tsai CY, Chou HK, Chen CC, Liou SW. Refractive change after pars plana vitrectomy. *Taiwan J Ophthalmol* 2012;2(1):18-21

21 Mohney BG. Axial myopia associated with dense vitreous hemorrhage of the neonate. *J AAPOS* 2002;6(6):348-353