Pars plana vitrectomy with/without encircling scleral band for treatment of retinal detachment associated with buphthalmos

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Abstract
Background: To evaluate the results of pars plana vitrectomy (PPV) and silicone oil (SO) tamponade with or without encircling band for repair of rhegmatogenous retinal detachment (RRD) in children with buphthalmos.

Patients and methods: Retrospective comparative nonrandomized interventional case series including consecutive patients who underwent PPV with or without encircling band and SO tamponade for RRD associated with buphthalmos.

Results: The study included 19 eyes of 19 children. Mean age was 8 years, range 3–16 years. Mean follow-up period was 28 months, range 19–63 months. Globe survival has been achieved in 15 out of 19 eyes (79%). Phthisis bulbi was reported in four cases (22%). Eight patients (42%) achieved ambulatory vision. Most eyes initially achieved anatomical success.

Conclusion: Despite the poor visual and anatomical results of RRD repair in eyes with buphthalmos, globe survival might be the rationale for surgery in such cases. Globe preservation could avoid the psychological and social consequences of phthisis bulbi in non-operated children.

Keywords: Retinal detachment, Buphthalmos, Globe survival after vitrectomy for buphthalmos

Introduction
Rhegmatogenous retinal detachment (RRD) is a major cause of loss of vision in adults. RRD occurs often in children. The reported incidence is 3–12% of all cases. Rarely, RRD is associated with primary congenital glaucoma [1–6]. The clinical features, underlying causes, outcome of surgical procedure and visual prognosis differ from those of adults. Surgical treatment poses major challenge to the vitreoretinal surgeon because children are more prone to develop macula-off RRD, proliferative vitreoretinopathy (PVR) and late presentation, hence longstanding retinal detachment. Consequently, the overall prognosis of surgery, particularly the visual outcome is highly-guarded [6–10]. Small-gauge PPV along with improved insight into the pathogenesis of RRD have considerably boosted the surgical outcome in these complicated cases [7–11]. Globe survival in this category of RRD might be a significant end-point for surgical intervention by preserving the globe and by avoiding sequelae of phthisis bulbi in childhood, particularly if patients needed an artificial implant for cosmesis [6–14]. The aim of the present study is to evaluate the results of PPV for repair of RRD associated with buphthalmos in children.
Patients and methods
This is a retrospective comparative interventional non-randomized case series that included children who had PPV for RRD associated with buphthalmos. Data of patients was retrieved from the medical records of all pediatric patients operated for RRD in a tertiary retinal center; Magrabi eye hospital, Tanta, Egypt, during the period from June 2008 to April 2019. Preoperative examination included; best-corrected visual acuity (BCVA) assessment and intraocular pressure (IOP) measurement when those were possible, slit-lamp examination, fundus examination using indirect ophthalmoscopy, and B-scan ultrasound evaluation.

Surgical technique
A single surgeon (HG) operated on all cases under general anesthesia. An encircling scleral band style 240 (2.5 mm) was combined with PPV except in patients who had functioning filtering bleb, aqueous filtering devices, e.g. Ahmed’s valve, or presence of scleral ectasia. Pars plana lensectomy was performed in all phakic children without intra-ocular lens (IOL) implantation. PPV consisted of core vitrectomy; triamcinolone acetonide (TA)-assisted induction of posterior vitreous detachment (PVD). Perfluorocarbon liquid (PFC) stabilized the posterior retina during shaving the vitreous base. The surgeon then performed air-fluid exchange, endo-laser retinopexy to retinal breaks and in an encircling 360° pattern. Finally, SO/air exchange was done. The surgeon routinely used SO 5000 centistokes (cSt) in all patients. Inferior surgical iridectomy (at 6 O’clock) was created in all aphakic eyes. Sclerotomies were closed using 7/0 vicryl sutures.

Postoperative care
Cooperative children were instructed to maintain prone position 8 h per day for 1 week. Those children had slit-lamp examination in the first postoperative day. If possible, fundus examination, and color fundus photography were done to assess and document retinal reattachment. The follow-up was scheduled at 1, 3, and 6 weeks, and then every 8 weeks. In each visit, we recorded BCVA and IOP when possible and assessed retinal reattachment. Uncooperative or young children had examination under general anesthesia; where anterior segment was examined using the surgical microscope, IOP was measured using schiotz tonometer and the fundus was examined using the indirect ophthalmoscope.

The term globe survival refers to eyes that did not need enucleation until the end of the follow-up period. Main outcome measures were retinal reattachment, improvement of BCVA and globe survival rate.

Management of recurrence
Patients who developed recurrent retinal detachment under SO during follow-up, had revision surgery that consisted of SO removal, TA-assisted removal of any residual vitreous cortex and peeling of epiretinal membranes. The surgeon performed relaxing retinotomy and/or retinectomy for immobile and shortened retinæ when needed followed by air/fluid exchange, drainage of subretinal fluid, endolaser retinopexy and finally SO 5000 cSt injection and closure of sclerotomies using 7/0 vicryl sutures.

Silicone oil removal
Silicone oil was removed in all eyes that had stable retinæ for at least 6 months. The technique consisted of 2-port 20-gauge or 23-gauge sclerotomies or both combined. One port was used for infusion, and the other for SO aspiration. The retina was thoroughly examined looking at the retinal periphery and the site of retinal breaks. SO was extracted manually or aspirated followed by multiple partial air/fluid exchange. Sclerotomies were closed using 7/0 vicryl sutures. The parents of all children recruited for the study provided a written informed consent after receiving thorough explanation of all surgical procedures entailed, prognosis for vision, and possible complications.

Results
Baseline characteristics (shown in Tables 1, 2)
The study recruited 19 eyes of 19 children. Male gender constituted 16 patients (84.2%). Mean age was 8.3 years, (range 3–16 years). At the time of recruitment, 18 children (94.7%) already had sub-scleral trabeculectomy (SST) and one child (5.3%) had glaucoma drainage device; Ahmed glaucoma valve done elsewhere. In terms of number and location of retinal breaks, 6 eyes (31.6%) had giant retinal tear (GRT), 3 eyes (15.8%) had multiple retinal tears; whereas 5 eyes (26.3%) had a single retinal break. We are unable to detect retinal breaks in 5 eyes (26.3%). An encircling scleral band was combined with PPV in 8 cases (42%). A scleral band was not applied in the remaining 11 cases. One eye had functioning Ahmed glaucoma valve, 6 eyes had scleral thinning and/or ectasia and 4 eyes had functioning bleb.

Post-operative anatomical outcome
All cases had successful retinal reattachment after the first surgery. During the follow-up period, 4 patients (21%) had revision of surgery and silicone reinjection due to recurrent RD. By the end of follow-up period 6 eyes (31.6%) had recurrent RD under SO that was either localized or total. SO was removed after stability of retinal
status for at least 6 months in 4 out of 19 cases (21%). SO was not removed in the remaining cases due to poor visual potential, hypotony or band shaped keratopathy, which prevented proper retinal evaluation. The rate of globe survival with free ocular movements in all directions of gaze was 78.9%.

Different degrees of exotropia developed in 6 eyes (31.6%). Four eyes (21%) developed phthisis bulbi at variable periods during follow-up.

Post-operative IOP
In 14 patients, IOP was controlled without anti-glaucoma drugs. IOP elevation developed after surgery in 5 patients, of which 2 patients required cyclophotocoagulation using diode laser and 3 patients had their IOP controlled using anti-glaucoma medications.

Corneal clarity
The cornea remained clear in 13 eyes. In 6 eyes SO migration to the anterior chamber with prolonged contact to the endothelium resulted in band keratopathy that contributed to their final poor visual outcome.

Post-operative functional outcome
Post-operatively, 8 eyes (42%) achieved ambulatory vision, of which 2 eyes had final BCVA of 2/60 and 6 eyes had final BCVA of 1/60. Final BCVA remained stationary (HM or PL) in 6 eyes (31.5%). Vision deteriorated in 5 eyes (26%). Poor visual outcome occurred mainly due to band keratopathy, PVR, and glaucomatous optic atrophy.

Discussion
Congenital glaucoma is a major cause of blindness in children, despite its low incidence (1:10,000 births). The disease is bilateral in approximately 75% of cases. Male patients account for approximately 65% of cases. Many authors corroborate the theory of polygenic pattern of inheritance [15]. In pediatric population, RRD associated with congenital glaucoma comprises a few number of children treated for RRD. Hence, there is limited data for this group of patients in published literature. Gupta et al.

Table 1 Data of patients who underwent an encircling scleral band with PPV

| Patients | Age (years) | Pre-operative visual acuity | Lens status | Retinal tear | Final retinal status | Corneal status | SOR | Postoperative visual acuity |
|----------|-------------|----------------------------|-------------|--------------|---------------------|---------------|-----|--------------------------|
| 1        | 9           | HM                         | Pseudophakic| GT           | Subtotal RD         | Band keratopathy | No  | HM                       |
| 2        | 9           | PL                         | Pseudophakic| GT           | Attached            | Clear          | No  | 2/60                     |
| 3        | 6           | HM                         | Phakic      | Undetected   | Total RD            | Band keratopathy | No  | NPL                      |
| 4        | 7           | HM                         | Pseudophakic| superior tear| Attached           | Clear          | Yes | 1/60                     |
| 5        | 9           | PL                         | Phakic      | superior tear| Total RD           | Band keratopathy | No  | NPL                      |
| 6        | 5           | HM                         | Pseudophakic| superior tear| Attached           | Clear          | Yes | 1/60                     |
| 7        | 4           | HM                         | Pseudophakic| Undetected   | Attached           | Clear          | No  | HM                       |
| 8        | 7           | HM                         | Pseudophakic|              | Attached           | Clear          | Yes | 1/60                     |

Table 2 Data of patients who underwent PPV without an encircling scleral band

| Patients | Age (years) | Preoperative visual acuity | Lens status | Retinal tear | Final retinal status | SOR | Corneal status | Postoperative visual acuity |
|----------|-------------|----------------------------|-------------|--------------|---------------------|-----|---------------|--------------------------|
| 1        | 10          | PL                         | Phakic      | Multiple     | Attached            | No  | Clear         | NPL                      |
| 2        | 8           | PL                         | Phakic      | GT           | Attached            | No  | Clear         | HM                       |
| 3        | 6           | HM                         | Cataractous | Undetected   | Attached            | No  | Clear         | 1/60                     |
| 4        | 16          | PL                         | Cataractous | Single       | Attached            | No  | Clear         | PL                       |
| 5        | 10          | HM                         | Aphakic     | GT           | Attached            | No  | Clear         | 1/60                     |
| 6        | 11          | HM                         | Cataractous | Multiple     | Attached            | Yes | Clear         | HM                       |
| 7        | 4           | PL                         | Pseudophakic| Undetected   | Total RD            | No  | Band keratopathy | NPL                      |
| 8        | 3           | HM                         | Phakic      | GT           | Attached            | No  | Clear         | HM                       |
| 9        | 10          | CF                         | Phakic      | Multiple     | Attached            | No  | Clear         | 2/60                     |
| 10       | 9           | PL                         | Subluxated  | Single       | Subtotal RD         | No  | Band keratopathy | NPL                      |
| 11       | 4           | HM                         | Pseudophakic| Undetected   | Attached            | No  | Clear         | 1/60                     |

GT giant tear, HM hand movement, PL perception of light, NPL no perception of light, RD retinal detachment, SOR silicone oil removal

CF counting fingers, GT giant tear, HM hand movement, PL perception of light, NPL no perception of light, RD retinal detachment, SOR silicone oil removal
reported the higher incidence of peripheral retinal degenerations and recommended screening especially in eyes with axial length ≥ 26 mm. Many authors reported poor visual outcome in cases of RRD associated with buphthalmos, despite retinal reattachment [6, 12, 13]. Satofuka and his colleagues reported poor visual outcome after management of RRD associated with buphthalmos in three eyes of three consecutive patients, despite retinal reattachment in all cases after PPV. In comparison, in the present study 8 eyes (42%) achieved ambulatory vision. Al-Harthi et al. [13] retrospectively reviewed 34 cases of present study 8 eyes (42%) achieved ambulatory vision. In three eyes of three consecutive patients, despite retinal reattachment in all cases after PPV. In comparison, in the present study 8 eyes (42%) achieved ambulatory vision. The authors reported retinal re-attachment in 3 eyes only for 6 months or longer and anatomical reattachment with visual improvement in 2 of the 5 eyes which had SO injection [12]. The results and conclusions of that study corroborate our rationale for this surgery. An important limitation of the study is the small sample size recruited due to the rarity of the condition studied. Another limitation is the retrospective nature of the study that relied on data collection from medical records from different glaucoma clinics with subsequent discrepancies in data compilation.

Conclusion
Repair of retinal detachment in eyes with buphthalmos carries poor visual and anatomical results, thus globe preservation might be the rationale of surgery in such cases. Surgical repair and globe preservation could avoid the psychological and social consequences of phthisis bulbi in non-operated eyes. Moreover, ambulatory vision might be obtained in some eyes.

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Authors’ contributions
HG conceptualization of the manuscript and the surgery of all cases. MH and HM the acquisition of data. SE, MH and AZ the analysis of data. KM, SE and AZ interpreted the data. MH, AG and ML data curation, methodology and validation. AG drafted the work. ML and HM substantively revised it. All authors read and approved the final manuscript.

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Availability of data and materials
All the study data and relevant materials are available on reasonable request from the corresponding author.

Declarations
Ethics approval and consent to participate
None of the authors has financial interests or other relationships including grants or research support relative to the content of the study. The institutional review board of Magrabi Eye Hospitals and Centers approved the present study. All parents or legal custodians of the children recruited in the study received through explanation of the nature of the surgical procedures entailed, the expected outcome and possible complications, and provided an informed consent. The study adhered strictly to the tenets of the Declaration of Helsinki (2013 revision).

Consent to publish
All the parents or legal custodians approved through a written consent to publish the results of the study.

Competing interests
The authors declare that they have no competing interests.

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