ABSTRACT

Purpose: To evaluate the long-term outcome of trabeculotomy-trabeculectomy as the primary surgical treatment for primary congenital glaucoma.

Methods: Twenty-two eyes of 14 consecutive patients with primary congenital glaucoma who underwent combined trabeculotomy-trabeculectomy as the initial procedure between 1981 and 2005 were selected for review. Records of ocular family history, age at onset of glaucoma, symptoms and signs, corneal integrity, intraocular pressure (IOP), gonioscopy, surgery, postoperative complications, visual acuity, refractive error, endothelial microscopy, axial length, visual fields and final status of the optic nerve head were entered into a computer database. The outcome, in terms of IOP obtained, was evaluated using Kaplan-Meier survival analysis.

Results: Cumulative probabilities of success, after performing combined trabeculotomy-trabeculectomy as the initial operative procedure, were 95.5% after 12 months and 78.2% after 24 months, with this rate being maintained during 15 years of follow-up. Four eyes (18.1%) required more than one operation.

RESUMEN

Objetivo: Valorar los resultados quirúrgicos a largo plazo de la trabeculotomía-trabeculectomía combinada (TTC) como tratamiento preferente del glaucoma congénito primario.

Métodos: Se seleccionaron 22 ojos de 14 pacientes consecutivos con glaucoma congénito primario en los que se empleó la TTC como procedimiento inicial entre 1981 y 2005. Se introdujo en una base de datos la información relacionada con la historia familiar, edad de inicio del glaucoma, síntomas y signos, integridad corneal, presión intraocular (PIO), gonioscopia, cirugía, complicaciones postoperatorias, agudeza visual, defecto de refracción, microscopía endotelial, longitud axial, campos visuales y estado final del nervio óptico. El resultado de la presión intraocular fue evaluado usando el análisis de supervivencia Kaplan-Meier.

Resultados: Las probabilidades acumuladas de éxito después de realizar una TTC como procedimiento inicial fueron del 95.5% a los 12 meses y del 78,2% a los 24 meses, manteniéndose esta proporción durante 15 años de seguimiento. Cuatro ojos...
The expression primary congenital glaucoma (PCG) is applied only to those cases where an anatomical defect exists for the camerular angle, due to isolated anomalies of development, which complicate the release of aqueous humour (1). This is the most common variety of infantile glaucoma, demanding an early diagnosis and a relatively urgent surgical treatment, as this condition may cause severe functional and morphological sequels.

Although goniotomy has been traditionally considered as the main option, other surgical alternatives have been proposed, such as ab externo trabeculotomy, trabeculotomy-trabeculectomy, or conventional trabeculectomy.

Combined trabeculotomy-trabeculectomy (CTT), also called trabeculo-trabeculectomy, was described by Nicolás Belmonte for PCG in 1979 (2). Several other authors have later published satisfactory results with this procedure (3-8).

CTT is difficult to assess in western societies, due to its limited use, and to the low incidence of PCG (1:10,000 newborns). In fact, the effectiveness of this technique has been analyzed only in a handful of articles, with most studies corresponding to arab population samples, where the incidence of PCG is higher due to the high levels of consanguinity (1:2,500 newborns) (9).

CTT has been used during the last 25 years as the main choice for treatment, with apparently good initial results. A retrospective check was thus applied to its long term effectiveness, with a mean follow-up time of 8.9 years (follow-up time was under 5 years for all series published before).

INTRODUCTION

The expression primary congenital glaucoma (PCG) is applied only to those cases where an anatomical defect exists for the camerular angle, due to isolated anomalies of development, which complicate the release of aqueous humour (1). This is the most common variety of infantile glaucoma, demanding an early diagnosis and a relatively urgent surgical treatment, as this condition may cause severe functional and morphological sequels.

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CTT has been used during the last 25 years as the main choice for treatment, with apparently good initial results. A retrospective check was thus applied to its long term effectiveness, with a mean (18,1%) requirieron más de un procedimiento debido a una PIO elevada persistente. Las complicaciones postoperatorias fueron raras. Al final del seguimiento, de 12 ojos de 8 pacientes cooperadores, la mejor agudeza visual corregida fue igual o mayor de 0,5 en el 75% (9 ojos).

Conclusiones: La TTC como cirugía primaria ofrece una alta eficacia a largo plazo en el control de la PIO sin tratamiento farmacológico, mostrando mínimas complicaciones quirúrgicas y una baja incidencia de reintervenciones.

Palabras clave: Trabeculotomía-trabeculectomía, glaucoma congénito primario, buphthalmos, glaucoma pediátrico, glaucoma infantil, glaucoma.

SUBJECTS, MATERIAL, AND METHODS

A total of 22 eyes were reviewed (designated I to 22), from 14 consecutive patients (designated I to XIV). All of them were diagnosed and operated on at the Alicante University Hospital between January 1981 and December 2005. All pre surgical and post surgical information and long term surgical results were assessed. All operations were performed by the same surgeon (JBM).

Since the number of alternative procedures was not large enough to allow for statistical analysis by comparison between different techniques, patients subject to other surgical techniques as initial treatments were excluded. Patients were also excluded for cases where paediatric glaucomas were associated with other ocular conditions, or systemic diseases.

A database was created including details on family history, age of onset of glaucoma, signs and symptoms, corneal integrity, intraocular pressure (IOP), gonioscopy, surgery, post surgical complications, visual acuity, refractive defect, endothelial microscopy, axial length, visual fields, final state of the papilla, follow-up, and end result.

Pre-operative diagnosis was based upon early signs and symptoms (photophobia, blepharospasms, tearing, corneal edema, and increase in corneal
size). Intraocular pressure was measured using Perkin's flatness tonometer under halothane-induced anaesthesia. Measurements were taken as soon as children were ready under the effects of the anaesthetic (within the first 10 minutes after induction, and before tracheal intubation).

Measurements of intraocular pressure (IOP) under anaesthesia were performed 6 months after surgery, and once a year after that. The eldest children and those who collaborated best were examined using a Goldmann tonometer.

The diagnostic criterion for surgical failure was established when additional surgery was required, due to intraocular pressure values above 21 mmHg (even under topical medication) or for IOP values above 16 mm Hg under general anaesthesia with persisting restraints, such as symptoms (photophobia, tearing) and signs (buphthalmos, cup progression, or optic nerve atrophy). Cumulative probabilities of success were determined in accordance with Kaplan-Meier survival analysis (SPSS v.14, SPSS Inc., Chicago, Illinois, USA).

No complementary tests were carried out due to the short age of some of the patients. When possible, depending on age and patient’s collaboration, assessments were made of refractive defect, biomicroscopy, state of the optic nerve, endothelial microscopy (SP 2000, Topcon, Tokyo, Japan), axial length (Ocuscan, Alcon, Irvine, CA, USA), and visual field (Humphrey, Zeiss, Dublin, CA). Up to 8 patients (12 eyes) collaborated enough to allow for reliable examinations to be performed.

**Surgical Technique**

The operations were started under general anaesthesia, with a superior rectus stitch with 4/0s silk suture in order to facilitate exposure of the upper conjunctiva. A fornix-based conjunctival flap was then created onto the upper limbus (at I or at XI). A rectangular tapete was then dissected towards the limb with an approximate size of 2 x 3 mm, into half or two thirds of the scleral thickness.

Using a high magnification, a radial incision was performed on the underlying sclera, using the blunt end of a slit knife (Fig. 1). The incision was carried out on the transition area between the ‘blue’ cornea (trabecular band) and the scleral tissue where Schlemm’s canal was theoretically located. The canal was identified when a reflux of aqueous humour was noticed as coming from the incision, occasionally mixed with blood.

It may sometimes be difficult to detect Schlemm’s canal when an abnormal anatomical elongation of the limbus or a thinned down sclera in buphthalmic eyes are present.

Vannas scissors are introduced at the incision level in the exposed canal, cutting 1 mm on either side. The double trabeculotome, right- and left (Sourdille-Paufique, Moria®) was then inserted into the incision, checking for obstacles to the advancement into the canal lumen. A controlled rotation was then performed towards the anterior chamber, thereby crossing the internal side of Schlemm’s canal, breaking the trabecular mesh and the angle’s embryonic tissue (fig. 2), whilst avoiding damage to other intraocular structures. In a similar way, the trabeculotome was then introduced through the other end of the incision (fig. 3). The internal side of Schlemm’s canal was finally cut in an extension of approximately 120º.

A small portion of tissue was subsequently extracted from the corneo-scleral bed, with an approximate size of 1 x 2 mm, and including some trabecular mesh and a portion of Schlemm’s canal (trabeculectomy), followed by a peripheral iridectomy (fig. 4). The scleral tapete was then repositioned by performing 2 interrupted stitches with 10/0 nylon suture onto the corners, and then burying the stitches. The conjunctiva was closed using interrupted or 8/0 continuous reabsorbable sutures.

**Fig. 1: A radial incision is carefully performed using a knife on the transition area between the ‘blue’ cornea (trabecular band) and the scleral tissue, in order to identify Schlemm canal.**
Antibiotics and corticoids were applied after surgery. Eyes were occluded for unilateral cases, and omitted for bilateral occurrences. Post surgical treatment was dispensed for three to four weeks, reducing prescription in accordance with clinical appearance and evolution.

RESULTS

22 eyes were analyzed for 14 patients, 10 males (71.4%) and 4 females (28.5%). The disease was bilateral in 8 children (57.1%) and unilateral in 6 (42.8%). A family history of PCG was present for only one case. In unilateral cases the right eye was involved in 66.6% of cases, with the left eye compromised in the other third.

Age at the time of diagnosis varied between 1 day and 23.5 months (mean value 2.3 months). Suspicion of congenital glaucoma was based on increased corneal size (22 eyes; 100%), corneal edema (14 eyes; 63.6%), tearing (12 eyes; 54.5%), photophobia and blepharospasmus (13 eyes; 59%). The mean horizontal corneal diameter measured was $13.4 \pm 1.1$ mm (range from 12 to 15 mm). Mean values for pre surgical IOP before first surgery was $20.1 \pm 4.2$ mm Hg. Follow-up time extended from 1 to 24 years, with a mean value of 8.9 years (118.8 months). Nystagmus by fixation was seen for one of the children.

The cumulative probability of success for IOP with one procedure only was 95.5% at 12 months, and 78.2% at 24. This proportion was constant during the 15 year follow-up. However, only 5 patients (8 eyes) reached 10 years of follow-up. Most surgical failures occurred during the first two years (fig. 5).

Four eyes (18.1%) required more than one operation due to high IOP values, despite the topical hypotensive treatment applied. The surgical technique selected for all re-operations was trabeculectomy. After a second operation IOP went back to normal levels in 2 eyes. One of these needed additional topical betablockers. On the 2 eyes remaining for the same patient (patient I) ocular tension was
surgery was performed, with poor functional results. The eye was then involved in an accidental trauma with corneal laceration and hyphema, which was resolved spontaneously within a few days, with ocular pressure under control, but with total loss of vision.

Only 3/22 eyes (10.9%) showed severe complications with an unfavourable final evolution, with two of those eyes for the same patient.

Mean IOP in the last measurement under general anaesthesia was 10.89 ± 4.1 mm Hg, with a mean reduction of 10.82 ± 4.36 mm Hg. IOP was under control for 20 eyes (90.9%) at the end of follow-up, after one or two interventions, with only one eye requiring additional topical hypotensive treatment. All blisters were either diffuse or slightly elevated for all eyes with IOP under control.

No variations were noticed for the horizontal diameter of the corneas (mean 13.4 ± 1.3 mm), as compared with pre surgical measurements. A transparent cornea was noticed at the end of follow-up for 13 eyes (59.05%), with 5 eyes (22.7%) presenting a mild corneal opacity, with acceptable transparency, and 2 eyes (9.09%) showing a persistent diffuse opacity. The two corneas of patient I (9.09%) mentioned above could not be assessed, for obvious reasons.

Criteria were met by the eyes of 8 collaborating patients for studying visual acuity, biomicroscopy, IOP (as measured with Goldmann tonometer), endothelial microscopy, axial length, and campimetry. Patient I was excluded from the analysis, despite his willingness to collaborate. Table I below shows a summary of the exploration results.

The best visual acuity with correction was equal or better than 0.5 for 9 eyes (75.0%), and under 0.1 for 2 eyes (16.6%). Loss of vision was attributed mainly to corneal opacity or amblyopia. Ophthalmoscopy showed an increase in papillary excavation for 2 eyes. Reliable campimetric data was obtained for 4 eyes only, for which no significant campimetric alterations were visible.

**DISCUSSION**

Surgery is the only effective treatment for PCG. Goniotomy ab interno is the main choice for a majority of surgeons, when corneas are transparent enough to allow for adequate visualization of the camerular angle (10-13). On the other hand, Sch-
lemm’s canal is not sectioned during goniotomy, and no actions are performed involving certain histopathological alterations described for PCG, such as immaturity of the trabecular mesh or Schlemm’s canal (1,14,15). This could explain surgical failures in some cases, and the need for a second procedure.

CTT creates two exit ways for aqueous humour: the first path (trabeculotomy) sections the total thickness of the trabecular mesh, thus communicating the anterior chamber and Schlemm’s canal, whilst the second exit way (trabeculectomy) allows for the creation of a supplementary fistula for drainage between the anterior chamber and the subconjunctival space (5,7). This double exit path would account (at least partially) for the high success rates in IOP control with a single procedure (4,5).

Furthermore, CTT offers a significant advantage for those cases where visualization of the angle is constrained by corneal opacity (63.6% in our series), by acting on an area well under control for the anterior segment surgeon using the microscope.

The main drawbacks for this technique, however, include the technical difficulties related to dissecting a lamellar scleral flap in eyes heavily thinned out by buphthlamos, accurately locating and identifying Schlemm’s canal in order to introduce the trabeculotome in cases of extreme megalocornea, where the limbic area is heavily altered, and a ‘blind’ rupture is produced of Schlemm’s canal and the trabecular tissue, since no direct view of the angle is possible (as opposed to goniotomy). In any case, supplementary trabeculectomy provides an additional exit way for aqueous humour even when Schlemm’s canal or its canalización with the trabeculotome are not accurately performed, so that IOP may decrease by filtration. Finally, areas for possible future surgery in case of surgical failure are limited, due to involvement of the perilimbic conjunctiva.

The authors decided to select CTT as their main choice for opaque and transparent corneas due to its comprehensive microscopic control, familiarity with the surgical area, the good results obtained from the start with a single procedure, and the small proportion of complications.

The positive outcome of this analysis is supported mainly by excellent long-term surgical results (8.9 years on average), a low rate of new surgical interventions, good control of IOP, reduced signs and symptoms, and functional visual recovery. These results are all similar to those obtained in other studies, even with a much shorter follow-up period (3-8).

Complications for CTT are in practical terms not different from those recorded for each of the methods applied separately (7). Mild hyphema was specially prevalent, with no atalatia, or problems derived from the filtration blister (infection, thinned out blisters, perforation, etc.). Iris enclavamiento at the level of the trabeculectomy was significant for one eye only.

Most surgical failures occurred within the first two years, and new standard trabeculectomies were scheduled as a second procedure. The underlying nature of the disease and an early diagnosis have a strong influence on the evolution and the prognosis of all cases (16). The unfavourable final results for
three eyes (two for one patient alone) were possibly related to the nature of the malformation, and to a larger dilation in the primary surgical treatment. On the other hand, it seems clear that keeping IOP under control for these eyes was not enough to ensure good functional results. Attention must be paid to each patient individually, treating anisometropia or the likely amblyopia with an early optical correction, or eventually through occlusion exercises.

The retrospective design and the small number of patients were two of the limitations of the current study, which prevented the authors from reaching final conclusions derived from the information available. However, in our opinion, CTT is safe, effective, and predictable enough for it to be selected as the primary surgical treatment for PCG, at least when conventional goniotomy is either not properly mastered or it may become the source of additional problems.

REFERENCES