Prognosis of primary ab externo surgery for primary congenital glaucoma

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Abstract

Background—The strategy of pressure reducing surgery in primary congenital glaucoma has changed over the last decade. Ab externo filtering procedures—for example, trabeculotomy or trabeculotomy combined with trabeculectomy, have now been accepted even as primary intervention.

Methods—The authors reviewed 61 eyes in 35 consecutive patients with primary congenital glaucoma, who underwent different types of initial ab externo surgery between 1988 and 1996 (median follow up 36 months) to determine the efficacy of different surgical techniques and the influence of various risk factors.

Results—Trabeculotomy was performed in 17 eyes (27.9%), trabeculotomy with trabeculectomy in 15 eyes (24.6%), and trabeculectomy in 29 eyes (47.5%). Regarding age, preoperative intraocular pressure, corneal diameter, ocular axial length, and incidence of corneal haze the subgroups were comparable. Success rates of trabeculotomy, trabeculectomy, and a combined procedure did not significantly differ when assessed by life table analysis. Patient age under 3 months (p=0.014) and an ocular axial length of 24 mm or more (p=0.016) proved to be major risk factors for primary ab externo surgery failure. A second operation was necessary in 20 of 61 eyes (32.8%) during follow up.

Conclusion—Prognosis of primary ab externo glaucoma surgery in primary congenital glaucoma seems to be governed more by the individual course and severity of the disease than by modification of surgical techniques.
surgery for primary congenital glaucoma from the middle of 1988 to 1996; patients with a history of previous surgery, with complex ocular syndromes (for example, Axenfeld–Rieger syndrome, aniridia, Sturge–Weber syndrome) or with any suspicion of secondary glaucoma were excluded.

Diagnosis of primary congenital glaucoma was always established by an examination under general anaesthesia using halothane. Informed consent for pressure reducing surgery was obtained from the parents at least 1 day before examination under anaesthesia.

Indication for pressure reducing surgery was established if four or more of the following criteria were fulfilled: (1) typical symptoms (epiphora, photophobia, blepharospasm), (2) cloudy cornea, (3) increased intraocular pressure (>18 mm Hg by applanation or >23 mm Hg by indentation under deep general anaesthesia with halothane), (4) abnormally increased corneal diameter or ocular axial length in comparison with the growth curve of normal eyes, (5) if visible, a pathological excavation of the optic disc and dysgenetic signs of the chamber angle—for example, mesodermal remnants or fine iris roots, (6) primary congenital glaucoma of the contralateral eye.

Success of pressure reducing surgery was determined by an intraocular pressure <18 mm Hg under general anaesthesia or <21 mm Hg in the awake child measured by applanation, stable axial length, improvement or at least stability of the optic disc excavation, and clearing of the cornea. Our definition of success excluded any antiglaucomatous medical treatment. Visual function was not taken as a criterion, since the mean age of patients included in the study was too young to obtain reliable results concerning visual acuity or perimetry.

Choice of surgical approach depended on two particular factors: (1) the year in which surgery was performed (Fig 1), as primary trabeculotomy alone was not introduced until 1993, while trabeculotomy was the standard procedure in the late 1980s; (2) the intraoperative identification of Schlemm’s canal: in at least four eyes an intended trabeculotomy or combined procedure turned into a trabeculectomy because Schlemm’s canal could not be localised. Pressure reducing interventions were performed by different surgeons all experienced in the field of paediatric eye surgery under supervision of the same senior surgeon (GKK). All initial pressure reducing ab externo interventions took place under general anaesthesia. A limbus based conjunctival flap was dissected in one of the upper quadrants without excision of Tenon’s capsule. Subsequently, a lamellar rectangular scleral flap of 4 × 4 mm size was dissected crossing the grey-white borderline zone into the clear cornea. In the case of trabeculotomy a radial incision was done to find Schlemm’s canal. A McPherson trabeculotome was inserted into the lumen of Schlemm’s canal on either side of the incision, if possible, and carefully rotated into the anterior chamber with the other parallel arm as a guide. In the case of trabeculectomy, after reaching the grey-white border and limbal clear cornea a trabeculectomy piece of 1 × 3 mm size was excised with the diamond knife and a small peripheral iridectomy was performed. In the case of trabeculotomy with trabeculotomy, both procedures were conducted subsequently as described. Finally, the scleral flap was loosely refixed using two 10-0 nylon sutures. The conjunctiva was closed with a running 8-0 Vicryl suture. The anterior chamber was filled with a balanced salt solution and the blood was washed out. Surgery was completed by a subconjunctival injection of dexamethasone and methylcellulose, and the eye was dressed with atropine and antibiotic corticosteroid ointment. Postoperative treatment comprised topical administration of combined antibiotic corticosteroid medication for 7–10 days.

Follow up data were collected from patients’ charts, by direct contact with the external ophthalmologist and by voluntary re-examination in our department.

Statistical analysis was performed using the software program PRISM Version 2.0 (GraphPad Software, Inc, USA). For evaluation of the postoperative outcome, Kaplan–Meier survival curves for censored data were plotted, and significance levels determined using the log rank test. Differences of distribution for different risk factors between the subgroups treated by different ab externo techniques were assessed by means of the Kruskal–Wallis test.
Results

SURGICAL OUTCOME

Primary pressure reducing surgery in patients with newly diagnosed primary congenital glaucoma was performed in 61 eyes (32 right eyes, 29 left eyes) of 35 patients. Among the 35 patients with primary congenital glaucoma 21 (60%) were male and 14 (40%) were female. Unilateral glaucoma was found in nine patients (25.7%), and bilateral glaucoma in 26 (74.3%).

Primary trabeculotomy was performed in 17 eyes (27.9%) of 11 patients, primary trabeculectomy in 29 eyes (47.5%) of 17 patients, and primary combined trabeculotomy-trabeculectomy in 15 eyes (24.6%) of 10 patients. In 22 patients with bilateral glaucoma the same type of surgery was conducted in both eyes. Only in four patients were different techniques of surgery performed in the two eyes (combined procedure and trabeculotomy or trabeculectomy). Age at initial surgery and follow up with range of values are indicated in Table 1. The difference regarding age at initial surgery was not significant (p=0.54), while in follow up of the subgroups clear differences were found (p<0.001) owing to the fact that trabeculotomy only became a primary procedure in recent years (1993–6). Distribution of various clinical factors were similar for all surgical subgroups assessed by the Kruskal–Wallis test as indicated in Table 2.

Kaplan–Meier survival analysis (Fig 2) revealed no significant difference in the surgical outcome between trabeculotomy and trabeculectomy (log rank test; p=0.5), nor between the combined procedure and trabeculotomy (log rank test; p=0.37) or trabeculectomy (log rank test; p=0.25). Success rates after 6, 24, and 60 months are indicated in Table 3. During follow up 20 eyes (32.8%) of 14 patients required secondary pressure reducing surgery. Combined trabeculectomy-trabeculotomy was performed in one eye, the remaining cases were treated by trabeculectomy as secondary intervention. Secondary filtering procedures (Fig 3) showed a significantly higher failure rate than primary trabeculectomy (p=0.03).

Taking into consideration large axial length and corneal diameters, high intraocular pressure, and young age at time of surgery as independent risk factors for surgical failure, we studied the outcome in patients with a specific risk factor by life table analysis. Buphthalmic eyes in patients younger than 3 months (n=20, p=0.014) and with an axial length of >24 mm (n=12, p=0.016) actually had a significantly different outcome from the remaining eyes (Figs 4 and 5), while the outcome in patients with corneal diameter of >14 mm (n=14, p=0.44) and a preoperative intraocular pressure of >24 mm Hg under general anaesthesia (n=13, p=0.41) was not significantly different. Outcome in Turkish or Arabian patients with glaucomatous eyes who underwent pressure reducing surgery was not significantly worse (n=10; p=0.165) than that in white patients.

COMPLICATIONS

Chamber angle bleeding with small hyphaema occurred in 11 eyes (65%) during trabeculotomy, in four eyes (27%) during combined trabeculotomy-trabeculectomy, and in seven eyes (24%) during primary trabeculectomy. The blood was usually absorbed within the first postoperative day. Prolapse of ciliary processes was reported for two eyes during trabeculectomy (7%) and for one eye during the combined procedure (6%). Postoperative iris incarceration was reported in one eye following trabeculotomy (7%) and for one eye during the trabeculectomy (11%), but pressure reducing surgery was successful in each of the four eyes.

Severe postoperative complications (within 6 months after surgery) comprised subluxation of the crystalline lens in one eye following pri-
the psychological situation of initial diagnosis.

Moreover, the small number of patients leads to a limited statistical power of the study. On the other hand, primary congenital glaucoma predisposed to poor results for both procedures in general. Elder, who exclusively studied non-white people. Besides, the portion of patients manifesting glaucoma soon after birth was relatively high in this study and obviously predisposed to poor results for both procedures in general. Elder, who exclusively studied Palestinian patients younger than 1 year, reported better results for combined trabeculotomy-trabeculectomy procedures (93.5%) after 2 years compared with trabeculectomy; however, the incidence of corneal haze in the trabeculectomy group was obviously higher (82% versus 56%), whereas other preoperative factors (for example, age, intraocular pressure, corneal diameters) were similar in both groups. The author explained the superior outcome of the combined procedure by improving outflow through the trabecular meshwork as well as through the sclera. Although combined procedures likewise seemed to have a favourable outcome after 2 years in our study, the advantage of this procedure over trabeculectomy was far from statisti-
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et al

et al even observed a 5

year success rate of 92% after the first

trabeculectomy in 13 eyes with primary

glaucoma congenital, although no patients

younger than 4 months of life, nor patients

with a corneal diameter larger than 13.5 mm

were treated in this study.13 The rate of

re-

surgery (34%) after initial trabeculectomy

in the study of Detry-Morel and Feron was

similar to ours; they also underlined poor

prognosis for neonatal forms of congenital

glaucoma and highly myopic eyes.12

Although primary congenital glaucoma is
described as an entity with the leading

pathological feature of trabeculodysgenesis

resulting in pathologically increased outflow

obstruction,17 21 the starting point for pressure

reducing surgery varies considerably and the

prognosis of any surgery is thought to be influ-

enced by the individual nature of dysgenesis.5 14 15 25

Pronounced symptomatics of high intraocular pressure during the first days

of life in a neonate are a limiting factor for the

outcome of pressure reducing surgery in congenital glaucoma.3 12 14 21 Russell-Eggitt et al

also reported a bad outcome for goniotomy in very young patients with congenital glaucoma,

thereby confirming the importance of age for

individual prognosis whatever the pressure

reducing surgery.21

Any association with other ocular or sys-
temic abnormalities evidently worsens surgical

prognosis in congenital glaucoma.13 24 Axial

length of the eye is also a critical factor,

whereas corneal diameter was not found to

influence outcome in our study. Possibly,

because of the stretched limbal anatomy, the

reading of the corneal diameter by calipers is

not as precise as A-scan measurement of the

axial length. Lack of prognostic power for the

preoperative intraocular pressure should

mainly be attributed to measurement in deep

general anaesthesia.22 A massive decrease of

intraocular pressure (IOP) has been shown in

animal models after application of halothane.27

Consequently, it is important to consider that

normal intraocular pressure in children ranges

between 9 and 12 mm Hg under general

anaesthesia with halothane. IOP measure-

ments between 16 and 18 mm Hg might

already be an indication of glaucoma when

using halothane. Ketamine anaesthesia avoids

this considerable decrease of IOP, but can lead

transiently to a minor increase of IOP.28

Ketamine anaesthesia in young children was

abandoned in our department of anaesthesiol-

ogy owing to the relatively long lasting effects

of ketamine and the safety of short anaesthesia

with volatile halothane.

Although trabeculectomy is widely per-

formed by many ophthalmic surgeons, this

procedure remains technically difficult in cases

of congenital glaucoma as the limbal anatomy

is usually distorted and the sclera extrava-

darily thin.21 This can lead to inadvertent sclera

perforation during preparation of the scleral

flap. Another potential risk can be the prolapse

of vitreous or ciliary body processes through

the peripheral iridectomy into the trabeculec-

tomy opening, especially with a primarily

dislocated lens. Retinal detachment after filter-

ing surgery in two patients of our group clearly

demonstrated the high susceptibility to retinal

tears of the stretched buphthalmic eye. Rice

also reported several cases of retinal detach-

ment in his large series of 246 eyes with

glaucoma congenital after goniotomy;22 although

the postoperative risk for retinal tears

should be higher in filtering procedures

owing to frequent postoperative hypotony of the eye.

Considering clinical results after goniotomy it

seems reasonable to perform goniotomy or

similar pressure reducing ab interno surgery

(for example, endoscopically guided goni-

otomy or laser goniotomy)29 30 as a primary

intervention, because the conjunctiva is not

damaged and thus future surgery is not preju-

diced. In order to avoid the use of antimetabo-

lites for as long as possible, a surgical strategy

sparing the conjunctiva from scarring at an

young age can be preferred in primary con-

genital glaucoma, since future re-surgery has

always to be taken into account.31 Application

of mitomycin or fluorouracil in young children

with a long life expectancy is associated with

the risk of late toxicity and potential mutagen-

ity of these antimetabolites, even though the

filtering procedures combined with the use of

mitomycin have become more and more

routine in antiglaucomatous surgery.32–34 Be-

sides, rupture of thin walled blebs, wound

leakage, late endophthalmitis, and long stand-

ing hypotony might be further deleterious

complications of filtering procedures with

mitomycin.35 37 However, in refractory cases

the use of antimetabolites or other methods16
to prevent scarring is unavoidable where

goniosurgery and ab externo procedures with-

out antimetabolites have failed.

We conclude that the different options for ab

eexterno glaucoma surgery do not clearly influ-

ence the outcome of surgery in primary

genital glaucoma. Moreover, early manifesta-
tion and large ocular dimensions are the key

to a limited prognosis of any pressure reducing

surgery in primary congenital glaucoma. As

re-surgery is often inevitable in congenital

glaucoma owing to long life expectancy, a step-

wise surgical strategy has to be devised,
starting with ab interno surgery and proceeding to conventional ab externo procedures, before using antimetabolites and cyclodestructive methods in refractory cases.