THE MANAGEMENT OF GLAUCOMA IN NANOPHTHALMOS

BY *F. Phinizy Calhoun, Jr.*, MD

NANOPHTHALMOS (FROM "NANOS" MEANING "DWARF") IS THE TERM USED BY DUKE-Elder[1] to describe a pure type of microphthalmos in which a small but otherwise normal eye is found. This relatively rare condition represents an arrested development of the globe in all directions after the embryonic fissure has closed so that the eye is reduced in volume without the presence of other congenital anomalies. The condition is usually bilateral, and there is a strong hereditary factor as shown by reported pedigrees of either dominant or recessive inheritance.

Clinically, the eyes are deeply set, the palpebral fissures are narrow and the most important features are marked hyperopia, small corneas, shallow anterior chambers, occasional macular hypoplasia, and the frequent late occurrence of glaucoma.

Very little has been written about the form and behavior of the glaucoma that occurs in nanophthalmos or microphthalmos. In several reports[2,3,4,5,6] only brief mention is made of its occurrence and management. However, O'Grady[7] reported the pathological examination of both small hyperopic eyes of a 23-year-old male with intractable glaucoma in which multiple glaucoma operations failed.

In 1974, Brockhurst[8] described the posterior segment complications which occurred in five patients with nanophthalmos who had undergone glaucoma surgery. The glaucoma in these patients was described as acute or chronic angle-closure. Conventional glaucoma surgery was successful in controlling the glaucoma in most of the patients. Four eyes in Brockhurst's series required subsequent lens extraction. In all eyes (eight) in which the fundus could be visualized, he found varying degrees of peripheral choroidal elevation and nonrhegmatogenous retinal detachment which made its appearance either early or late after either glaucoma or lens surgery and comprised the entity of uveal effusion previously described by him.[9,10]

The present report was prompted by the author's experience in the

TABLE I: CLINICAL SIGNS OF NANOPHTHALMOS

1. Extreme hyperopia with generally good corrected vision
2. Small corneal diameter
3. Shallow anterior chamber, with "vesuvio" pupil
4. Gonioscopically "slit" or grade I angle
5. Wide pulse pressure on tonometry
6. Occasional macular hypoplasia
7. Tendency to glaucoma in middle age

TABLE II: CLINICAL CHARACTERISTICS OF GLAUCOMA OCCURRING IN NANOPHTHALMOS

1. Slow, painless and progressive elevation of intraocular pressure in middle age
2. Paradoxical response to medical treatment
3. Failure of conventional glaucoma surgery
4. Tendency to serious posterior segment complications following anterior segment surgery

clinical management of the glaucoma in several patients in which the medical and surgical control was so difficult that it was felt useful to analyze the special features of the glaucoma which occurs in nanophthalmos.

To this end, six patients with bilateral nanophthalmos were studied; nine of the twelve eyes had various stages of glaucoma. It was hoped that the relationship of nanophthalmos to glaucoma could be determined and that guide-lines might be established for the management of future cases.

CASE REPORTS

CASE 1

A 56-year-old housewife, had been very farsighted all her life but had no symptoms of glaucoma. Vision in the right eye was 20/20 −2 with a +15.00 sphere; vision in the left eye was fingers at two feet due to exotropia and amblyopia since childhood. Horizontal corneal diameters were 11.0 mm in each eye; anterior chamber depths were 2.45 mm in the right eye, and 2.45 mm in the left eye; applanation intraocular pressures were 19 mm Hg in the right eye and 18 mm Hg in the left eye with wide pulse pressure. Gonioscopy revealed a narrowed angle entrance with only a slit opening, even upon tilt and pressure. No peripheral anterior synechias were seen. A Mydriacil provocative test was negative. Tonography revealed a C factor of 0.28 and $P_{o/C}$ of 64 in the right eye; and a C factor of 0.17 and a $P_{o/C}$ of 100 in the left eye, indicating a facility of outflow which was normal in the right eye and borderline in the left eye. The optic discs were normal. Visual fields were normal in the right eye, and showed a small central scotoma in the left eye.

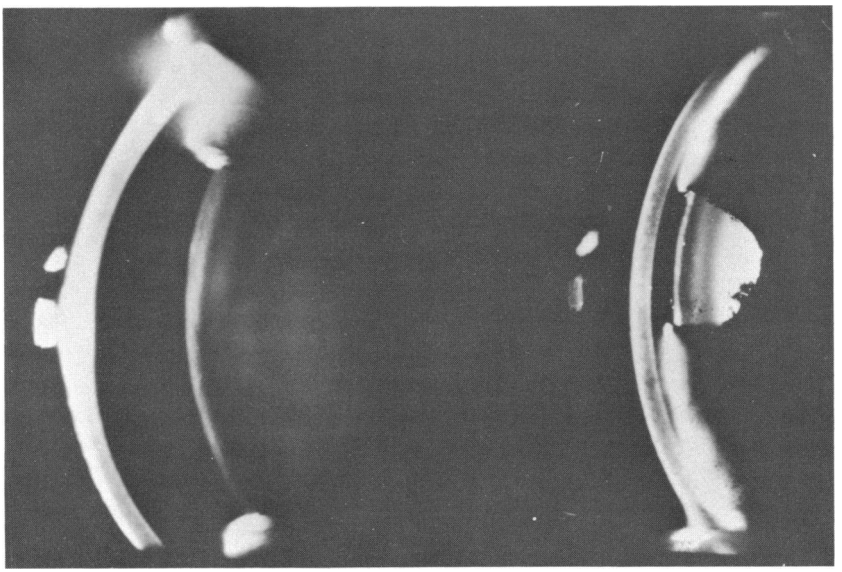


FIGURE 1
Case 2. Optical section of OD on the left and OS on the right. OD, shown with pupil dilated, had a pressure of 18 mm Hg, narrow angle and negative mydriatic provocative test. OS, with a pressure of 22 mm Hg, shown with very shallow chamber and cataract, after peripheral iridectomy and later filtering operation.

COMMENT

Although the corneal diameters and anterior chamber depths are only slightly reduced, these eyes are unusually short and hyperopic and must be considered to be nanophthalmic. Although the angles are theoretically not blockable at the present time, the patient is subject to acute or subacute angle-closure in the future. She is more apt to develop chronic angle-closure of the "creeping" type in which progressive peripheral anterior synechias might form in the recesses of the angle hidden from gonioscopic view by the slit angle entrance. Prophylactic miotics are contraindicated because of the possibility of producing relative pupillary block. If glaucoma develops, a peripheral iridectomy with tight closure should be performed following which miotics could be used if necessary.

CASE 2

A 42-year-old housewife, who had had poor vision and had worn strong glasses all her life, complained of recent blurred vision in her left eye and was found to have advanced glaucoma. The intraocular pressure was 18.4 mm Hg in the right eye, and 43.4 mm Hg (Schiøtz) in the left eye. Her corneas were small.

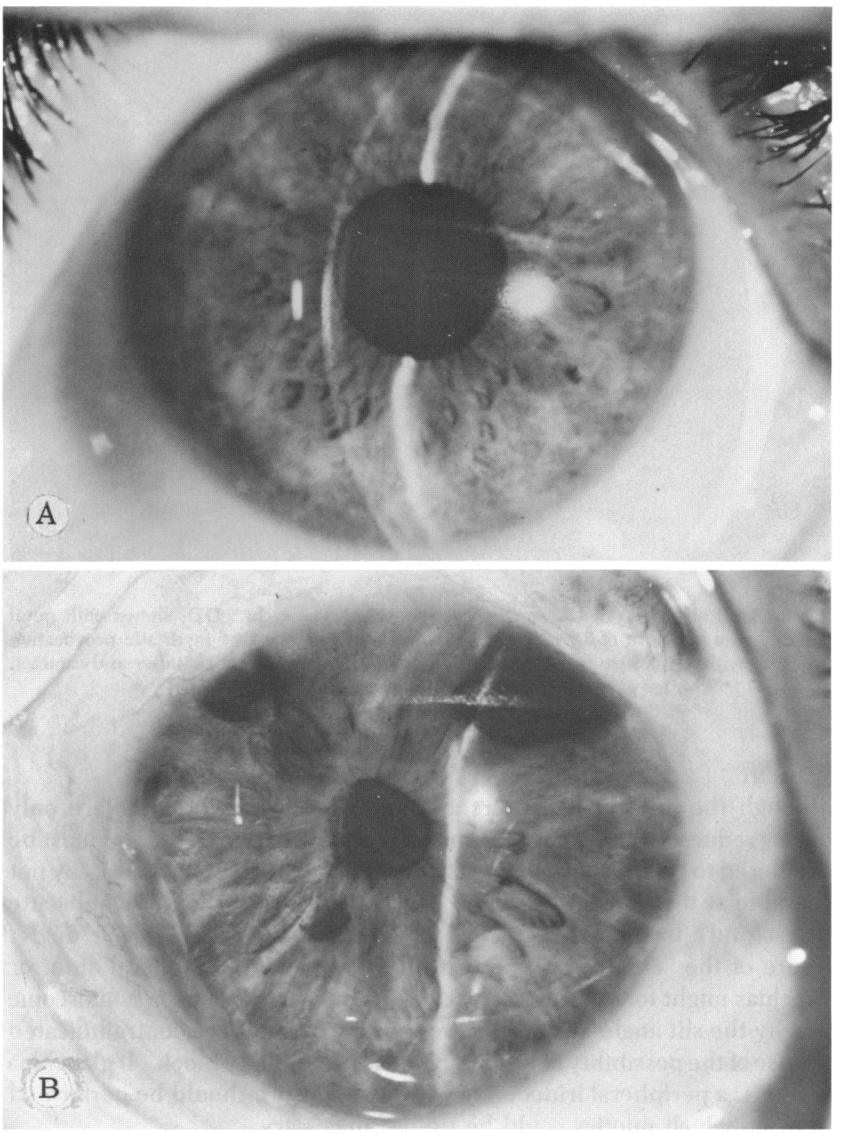


FIGURE 2

Case 2. Seven years after lens extraction OS. A: OD with corneal diameter 10.5 mm, anterior chamber depth 2.30 mm, narrow open angle, and pressure of 22 mm Hg. B: OS, with aphakic anterior chamber depth 3.50 mm, completely sealed angle, and pressure of 20 mm Hg.

The anterior chambers were very shallow and the chamber angle in the left eye appeared to be closed gonioscopically. Vision with her glasses was OD +14.75 sphere 20/70 and OS +12.00 sphere 20/200. The left optic disc showed glaucomatous cupping. Medical treatment failed to relieve the glaucoma in the left eye and a peripheral iridectomy was performed. This failed to benefit the glaucoma and an anterior lip sclerectomy was performed five months later. Although a small functioning bleb was produced, the intraocular pressure was not well controlled even with additional maximum medical therapy and she developed a cataract and reduction of vision to 20/400 in the left eye.

When seen by the author, the corrected vision was 20/70 in the right eye and light perception in the left eye (Figure 1). The corneal diameters were 10.5 mm in each eye. The anterior chambers were shallow. Gonioscopically, the chamber angle was open in the right eye even with dilatation, but the left appeared closed 360°. The intraocular pressure measured 18 mm in the right eye, and 22 mm in the left eye (by applanation). The lens in the right eye was clear; the left eye contained a nuclear cataract. The right fundus demonstrated a peculiar pale mottled appearance, however, no photographs were taken. The right visual field was normal.

Approximately sixteen months after the filtering operation an extracapsular lens extraction was performed on the left eye by her local ophthalmologist with resultant control of the intraocular pressure and vision of 20/70 with +21.00 sphere +2.50 Cyl ax 45°.

Seven years later (Figure 2), with treatment consisting only of 2% pilocarpine in the left eye twice daily, the vision with contact lenses was 20/80 in the right eye and 20/200 in the left eye. The intraocular applanation pressures were 22 mm Hg in the right eye and 20 mm Hg in the left eye. Horizontal corneal diameters were 10.5 mm in both eyes. Anterior chamber depth measurements were 2.30 mm in the right eye and 3.50 mm in the left eye. Gonioscopic examination showed a Grade I angle in the right eye with narrowed entrance and, by tilting, the meshwork and narrow ciliary body recess could be partially visualized without peripheral anterior synechias. The left eye showed a completely sealed angle. The right pupil was dilated without angle-closure revealing a clear lens and a peculiar pale mottled fundus with an absent foveal reflex. The left fundus could not be visualized.

COMMENT

This patient developed chronic angle-closure in the left eye. After an iridectomy and then a filtering operation failed, the glaucoma was controlled by a lens extraction. The cause of the long-standing amblyopia in each eye was not determined but the peculiar mottled pallor of the retina in the right eye suggested the possibility of macular hypoplasia.

CASE 3

A housewife had always been extremely farsighted, but had good corrected

vision. At the age of 31, she was told that she had glaucoma in the left eye and pilocarpine was used intermittently until the age of 44 when painless impairment of vision in the left eye prompted her to be examined by another ophthalmologist who found the following refractive error and vision: OD +11.00 sphere +1.50 cyl axis 100° 20/30, OS +12.00 sphere 20/80. The corneas were said to be small, the anterior chambers extremely shallow, and the intraocular pressure was 16.9 mm Hg in the right eye and 46.0 mm Hg in the left eye (Schiøtz). Gonioscopy was not performed at that time. The eye was not congested. The lenses were clear and the left disc showed moderate cupping. Maximal medical therapy reduced the pressure in the left eye only a few points. Five days later a clear lens extraction and sector iridectomy was performed on the left eye by the local ophthalmologist. Postoperatively there was transient choroidal detachment but the intraocular pressure remained controlled with mild glaucoma therapy and the vision five years later was +23.00 sphere +1.50 cyl ax 150° 20/100.

Ten years after the lens extraction the patient experienced a loss of vision in the left eye and a retinal surgeon described an almost total retinal detachment with peculiar choroidal pigmentation and deep pigmentation in the periphery. This gave the appearance of choroidal detachment. The right eye had similar choroidal pigmentary changes but no evidence of choroidal detachments. No retinal tears were found in the left eye but a dialysis of the ciliary pigment epithelium was seen in the lower nasal quadrant. A scleral buckling procedure was performed and although subretinal fluid did drain, a moderate subretinal hemorrhage developed. The presence of choroidal detachments could not be confirmed. At the time of operation the corneal diameters were 10.5 mm, the diameter of the globe was 15 mm and the ora serrata appeared to lie only one or two mm from the limbus in each eye. Postoperatively the subretinal fluid recurred and because the condition took on the appearance of an inflammatory detachment, massive doses of systemic steroids were given for two months, resulting in a gradual settling of the retina and choroid and restoration of vision to 20/100.

Meanwhile, the right eye which had been treated with miotics as a preventive measure began to show progressive elevations of pressure, beginning about five years following lens extraction on the left eye. On at least three occasions there were acute episodes of pain, congestion, and extremely high intraocular pressures. Following the subsidence of such an attack, the pressure was 17 mm Hg (Schiøtz) and gonioscopy showed the angle to be completely closed except for a small area below and temporally. A clear lens extraction and sector iridectomy was finally performed on the right eye by the same ophthalmologist three years before the retina operation on the left eye, and the glaucoma has been easily controlled since then (Figure 3).

At the present writing, seven years after lens extraction in the right eye and four years after the retina operation in the left eye, examination reveals the following: Vision OD +21.00 sph +1.00 cyl ax 90° 20/30+ and OS +23.00 sph 20/400. Corneal diameters are 11.00 mm (micrometer). Applanation intraocular pressures are 9 mm Hg in the right eye and 8 mm Hg in the left eye. Glaucoma treatment is being continued in the right eye. The fundi are as described

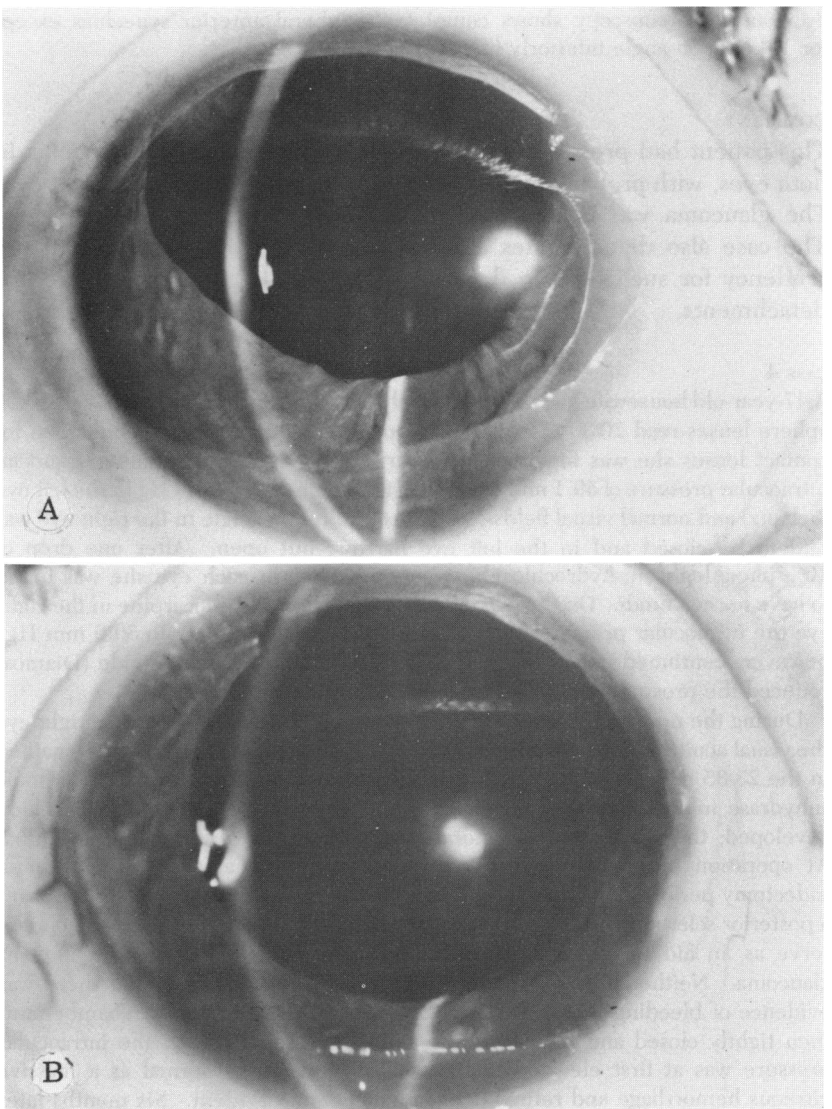


FIGURE 3

Case 3. Showing appearance of eyes after clear lens extraction and sector iridectomy performed seven years previously in OD. A: Fourteen years previously in OS. B: Corrected postoperative vision OD 20/30, OS 20/400, and pressure is OD 9 mm Hg and OS 8 mm Hg.

previously. Gonioscopy shows complete peripheral anterior synechias except for 10° of open-angle inferiorly in both eyes.

COMMENT

This patient had progressive painless chronic angle-closure glaucoma in both eyes, with probable episodes of acute angle-closure in her right eye. The glaucoma was controlled by clear lens extractions in both eyes. The case also demonstrates abnormal choroidal pigmentation and the tendency for such eyes to develop postoperative choroidal and retinal detachments.

CASE 4

A 47-year-old housewife had been extremely hyperopic all her life and with +16.00 sphere lenses read 20/30 in each eye. During the course of an examination for contact lenses she was found to have very shallow anterior chambers, and an intraocular pressure of 59.1 mm Hg in the right eye and 20.6 mm Hg in the left eye (Schiøtz) and normal visual fields. Gonioscopically, the angle in the right eye was said to be closed and in the left eye narrow, but open. After one drop of 10% phenylephrine hydrochloride (Neosynephrine) in each eye she was found to have normal fundi. On the day following the use of 2% pilocarpine in the right eye the intraocular pressure of the right eye was reduced only to 50.9 mm Hg, however, continued miotic therapy and the addition of acetazolamide (Diamox) reduced the pressure in the right eye to 26.6 mm Hg (Schiøtz).

During the next four years under maximum medical treatment to the right eye the visual acuity and field remained normal but the intraocular pressure remained in the 23-35 mm Hg range (applanation) with a wide pulse pressure. Carbonic anhydrase inhibitors had to be discontinued because of an idiosyncrasy which developed; the visual field deteriorated rapidly and severely in nine months. At operation the shallow anterior chamber was evacuated and a peripheral iridectomy performed. Since scleral indentation showed the eye to still be firm, a posterior sclerotomy was performed with the hope of reducing vitreous volume, serve as an aid to reforming the anterior chamber, and prevent malignant glaucoma. Neither fluid nor solid vitreous could be withdrawn and there was evidence of bleeding into the choroid and vitreous. The anterior chamber was then tightly closed and reformed with saline. Postoperatively, the intraocular pressure was at first elevated but gradually returned to normal as a massive vitreous hemorrhage and retinal detachment became evident. Six months later the vitreous had cleared sufficiently to see an infero-temporal retinal detachment with fixed retinal folds, heavy vitreous membranes and vision reduced to hand movements temporally. This condition remained unchanged for two years, with the intraocular pressure being 16 mm Hg in the right eye and 18 mm Hg in the left eye (applanation). The retinal detachment in the right eye became complete, with dense white preretinal membranes, and then the intraocular pressure, became elevated in the left eye. For the next five years up to the present time

the intraocular pressure of the left eye has steadily climbed to the high 20's and 30's and seems little influenced by weak miotics and tolerable dosage of carbonic anhydrase inhibitors (Figure 4).

At the present writing, her visual acuity is light perception in the right eye, and 20/30 in the left eye. The intraocular pressure is 12 mm Hg in the right eye, and 28-38 mm Hg (applanation) in the left eye, with a wide pulse pressure, and the visual field is beginning to show early glaucomatous defects. Examination of the left fundus shows a small but normal disc and marked increase in the light reflex of the retinal arterioles. Other details of her examination are: horizontal corneal diameters 10.25 mm; a keratometer reading of OD 52.25×180°×52.25×90°, OS 53.00×150°×51.51×109°; anterior chamber depths of OD 2.45 mm (postoperative), OS 1.90 mm. Gonioscopy shows the angle in the right eye to be closed temporally and inferiorly and open superiorly and nasally; in the left eye to be closed superiorly and temporally with nasal quadrant grade I, inferior quadrant "slit."

COMMENT

Progressive chronic angle-closure in both eyes with retinal detachment developing in the right eye following vitreous hemorrhage produced by a posterior sclerotomy performed at the time of peripheral iridectomy. Medical treatment of the left eye is showing but little benefit in preventing a slow rise in base pressure and surgical treatment is being considered.

CASE 5

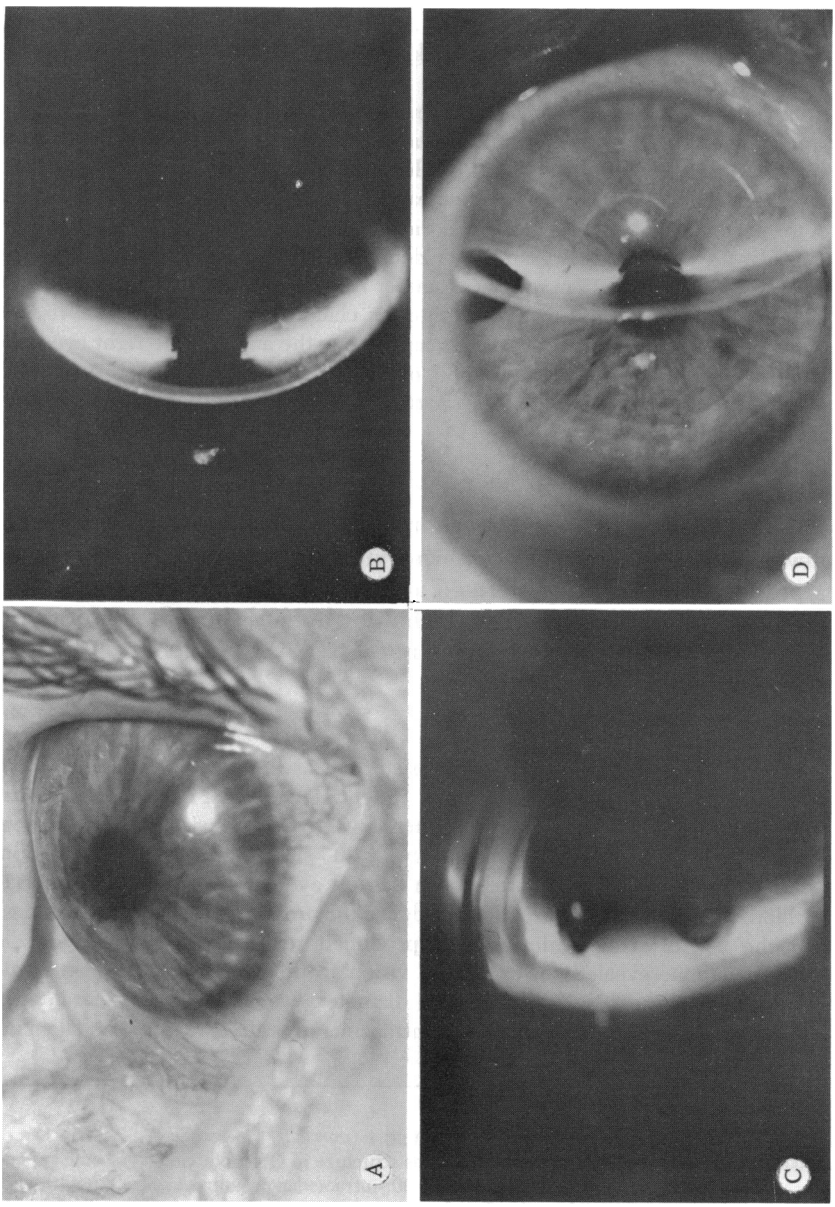
A 51-year-old housewife, and the older sister of Case 4 had always been very far-sighted but with good corrected vision. "Slight elevation" of intraocular pressure of the left eye was found but no treatment was given until six years later when, because of left sided headaches and eye pain, she returned for examination. At that time the intraocular pressure in the left eye was found to be 38 mm Hg (Schiøtz). Miotics and acetazolamide reduced the pressure to normal for a few days but the use of strong miotics then produced severe pain and congestion.

Examination revealed the corrected vision to be OD +15.25 sphere 20/40, OS +14.75 sphere 20/40. The corneal diameters were 11.00 mm, the anterior chambers shallow, and the applanation intraocular pressures were 22 mm Hg in the

---

FIGURE 4 *(overleaf)*

Case 4. A: Marked bowing forward of iris-lens structure in OS producing a volcanic appearance in the pupil ("vesuvio iris"). Vision 20/30, intraocular pressure 45 mm Hg with wide pulsation of tonometer and angle closed superiorly and temporally. B: Showing shallow anterior chamber measuring 1.90 mm in depth. C: Showing marked bowing of the iris upon gonioscopy. D: OD, seven years after peripheral iridectomy and posterior sclerotomy. Anterior chamber depth is now 2.45 mm, the pressure is normal without treatment, but the eye is blind from vitreous hemorrhage and retinal detachment.


B
D
A
C

right eye, and 50 mm Hg in the left eye. Gonioscopy showed a grade I open angle in the right eye and an inferior closed angle in the left eye. The left eye was slightly congested but the pupil was miotic. Despite stronger miotics, oral glycerin and eventually intravenous mannitol she developed signs of acute angle-closure with high pressure, steamy cornea, congestion, and severe pain. A peripheral iridectomy with tight closure and chamber reformation was performed. It was noted that even when the eye was open, scleral indentation showed it to still be firm. Postoperatively, there was a transient hyphema and the pressure remained elevated. During the next few months, she received maximum medical treatment but the glaucoma was never controlled. Slit lamp examination through the surgical coloboma of the iris showed adequate separation of lens equator from the ciliary processes. She refused a filtering operation. Rubeosis of the iris and cataract developed and the anterior chamber finally became completely flat (Figure 5A, B).

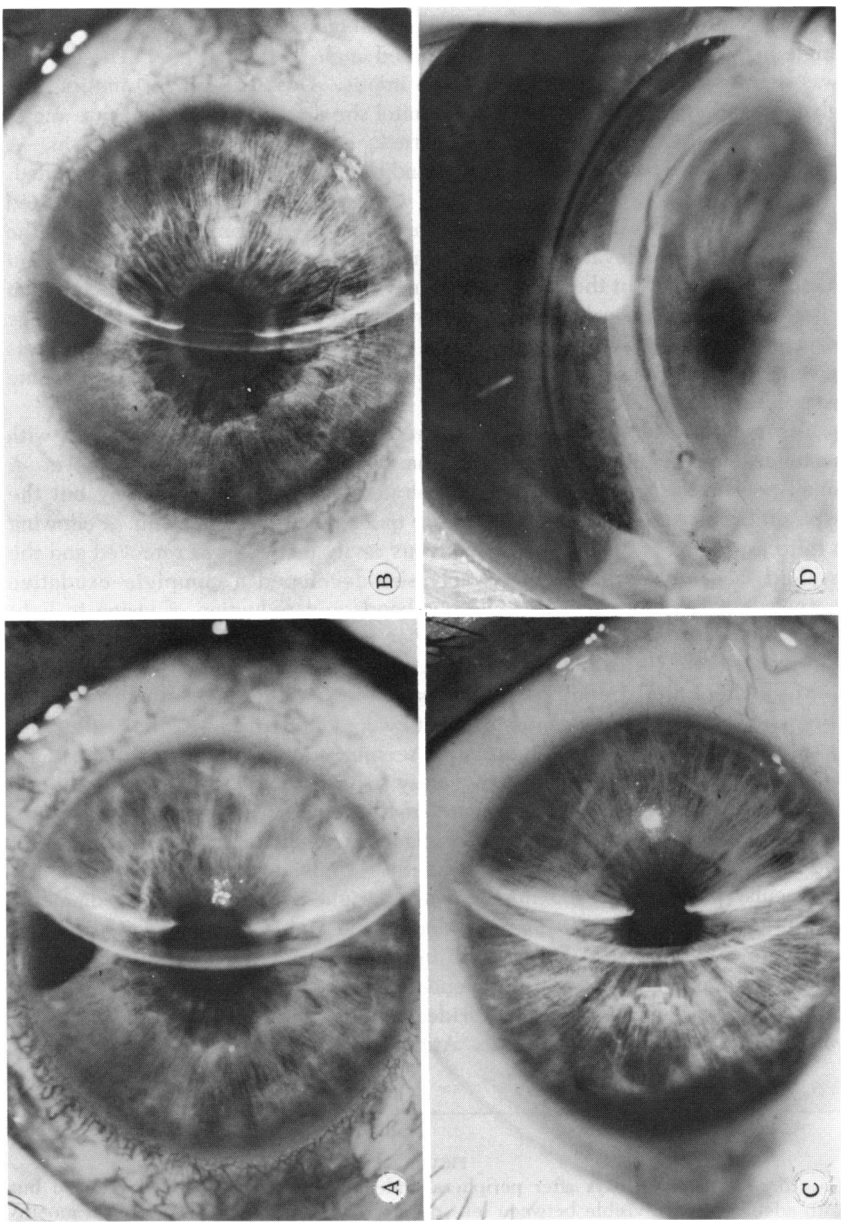
Ten months after the peripheral iridectomy an intracapsular extraction with sector iridectomy and posterior sclerotomy was performed on the left eye. A small pocket of subvitreal fluid was aspirated through the sclerotomy but the aspirant became blood-tinged and the eye quickly became firm again. Following a third aspiration of blood from the vitreous cavity the lens was removed and the hyaloid face incised. Postoperatively she developed a complete exudative retinal detachment with heavy vitreous bands and reduction of vision to light perception, however, the anterior chamber was deep and the intraocular pressure was 15 mm Hg without medication.

Meanwhile, the base intraocular pressure of the right eye continued to climb (Figure 5C, D). Accurate measurements were difficult due to the wide pulse pressure on tonometry. The pressure seemed unbenefitted or worsened by miotics and she developed an idiosyncrasy to all carbonic anhydrase inhibitors. Her vision and visual field remained unaffected for 3½ years but she then rapidly developed typical progressive glaucoma field defects and lessening of the hyperopia over a period of a few months. Under treatment her applanation intraocular pressure was 36 mm Hg in the right eye. The corneal diameter was 11.0 mm, the anterior chamber depth 1.50 mm, and gonioscopy revealed the angle to be closed 170° and upon inferiorly to one half of the trabeculum. Under general anesthesia and intravenous mannitol a trabeculectomy, an intraocular lens extraction, and a sector iridectomy were performed on the right eye without vitreous loss or hemorrhage. At the end of the operation, scleral indenta-

---

FIGURE 5 *(overleaf)*

Case 5. A: OS seven days after peripheral iridectomy with pressure not reduced but with adequate space visible between lens equator and ciliary processes. B: Ten months later after coninued miotic therapy showing loss of anterior chamber accompanied by iritis, posterior synechias, congestion, and high pressure. C: and D: OD prior to development of glaucoma, showing shallow anterior chamber, bowed iris, and narrowed angle entrance upon gonioscopy.


B
D
A
C

tion showed the eye to be firm. Immediate measurements of the rather small and globular lens were 8.5×8.0×4.5 mm.

At the time of the first postoperative dressing there was mild subconjunctival filtration, flat anterior chamber, and complete detachment of the retina, which was against the posterior iris surface (Figure 1A,B). It was thought that the patient had developed an acute massive choroidal effusion and retinal detachment following lens removal. The possibility of a giant retinal tear could not be ruled out at this time. Applanation intraocular pressure was 8 mm Hg. The patient was placed on massive systemic corticosteroids and 17 days later air injection and partial drainage of suprachoroidal fluid was performed. It was noted that the sclera was extremely thick at the drainage site 3.5 mm from the temporal limbus. Benefit from this procedure was only transient.

Under systemic corticosteroids the anterior chamber gradually reformed and the retinal balloon in the pupil receded temporally revealing massive pale choroidal detachments. The iris was adherent to the retina in a broad area temporally and a small area nasally. Intraocular pressures were 14 mm Hg in the right eye and 20 mm Hg in the left eye (applanation) with wide pulsations of the tonometry end point.

Five months later, the improvement from systemic corticosteroids seemed to have become stationary and the vision improved to finger counting at one foot. At this time careful lysis of the irido-retinal adhesions, incision of the hyaloid and bands of vitreous, unsutured drainage of suprachoroidal fluid nasally and temporally, and massive air injection were performed. The patient was kept in the prone position for three days, following which there was marked flattening of the choroidal and retinal detachments. With continued use of systemic corticosteroids the choroidal and nonrhegmatogenous retinal detachments gradually but incompletely settled and vision improved to finger counting seven months after the lens extraction (Figure 6C, D).

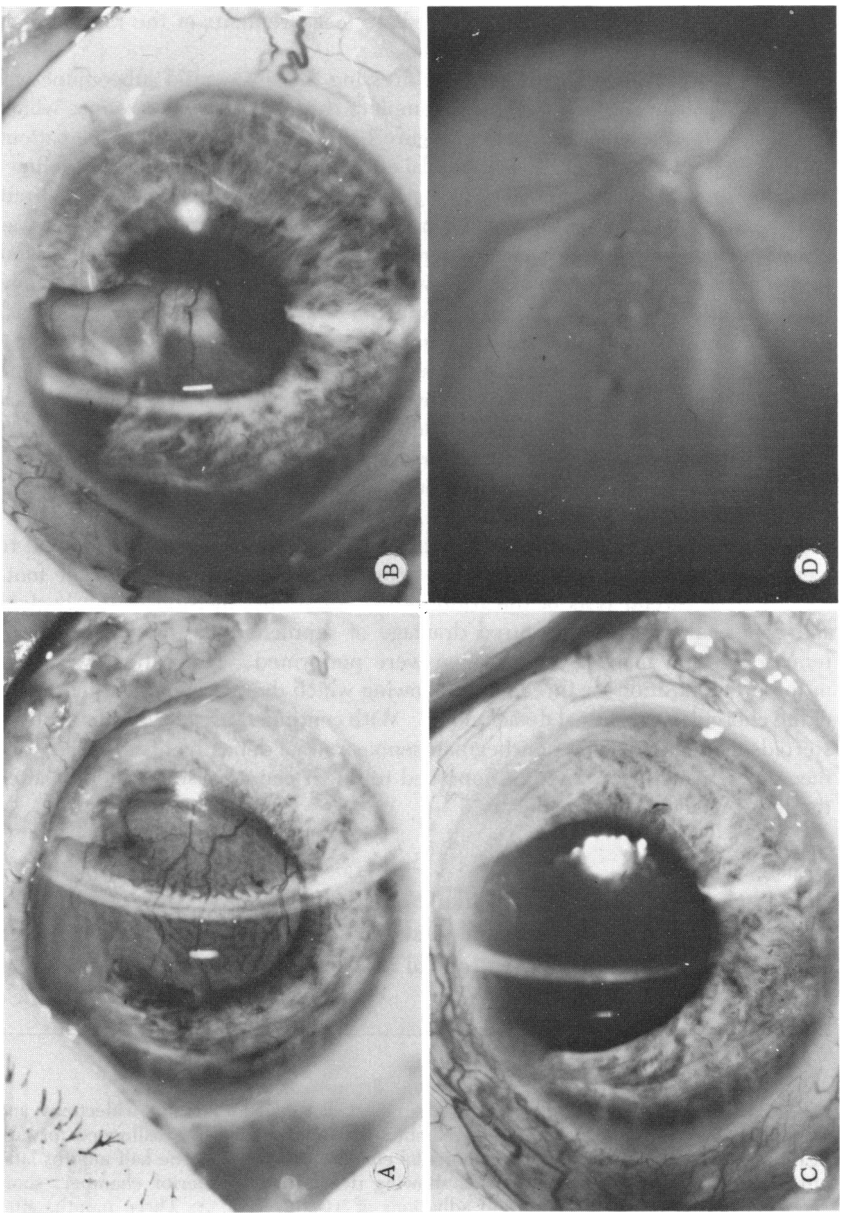
COMMENT

This patient behaved exactly like her sister (Case 4), except that the opposite eye developed glaucoma first and she demonstrated even greater allergies and idiosyncrasies to topical and oral medications. The course

---

FIGURE 6 *(overleaf)*

Case 5. OD A: Appearance three days after cataract extraction, sector iridectomy and trabeculectomy, showing flat anterior chamber with detached retina ballooned forward against iris, accompanied by massive choroidal effusion. B: Two and one half months later after treatment with systemic steroids showing deepening of anterior chamber, some retraction of the retinal balloon, and adhesions of iris to retina. C: Three months after lysis of irido-retinal adhesions and vitreous bands, drainage of suprachoroidal fluid, and massive air injection; showing clearing of pupil. D: appearance of the posterior pole with persistent flat nonrhegmatogenous retinal detachment but improvement of vision to 10/400.


B
D
A
C

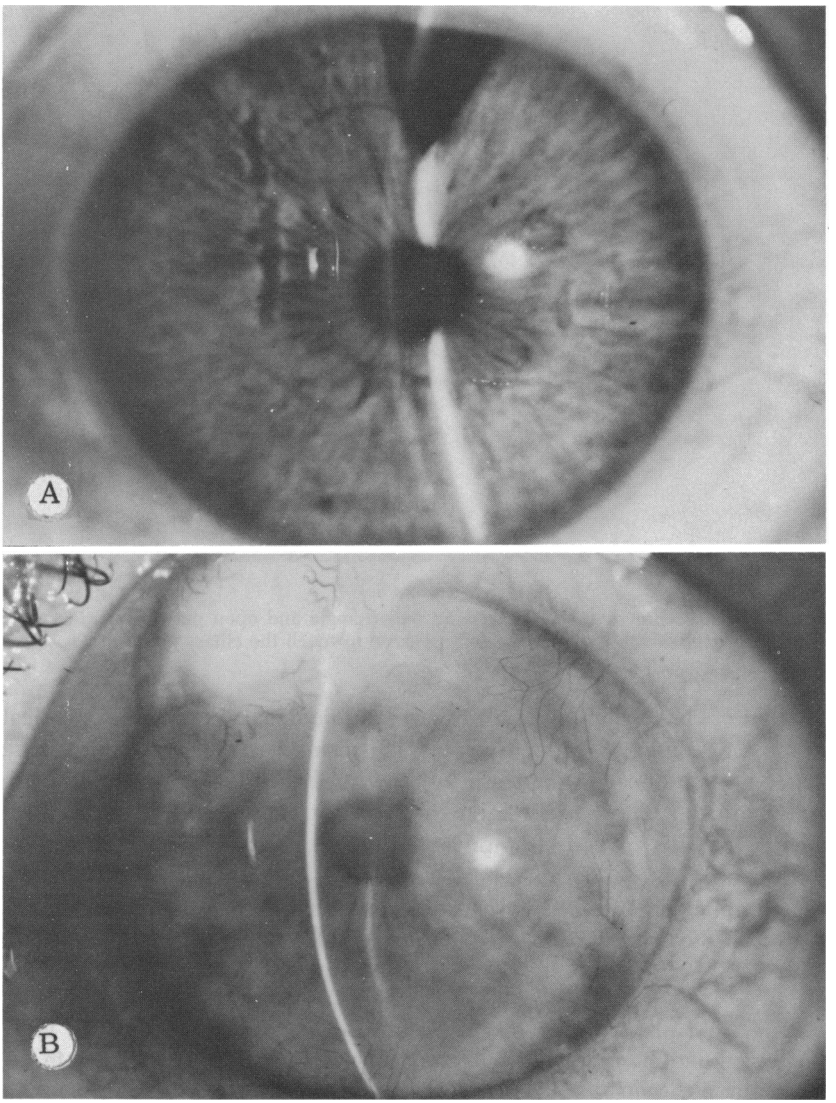


FIGURE 7

Case 6. A: OD and B: OS. Five years after peripheral iridectomy OD and peripheral iridectomy and posterior sclerotomy OS. Intraocular pressure OD remains at 28 mm Hg despite medical therapy, the anterior chamber depth is 2.30 mm, and the angle is sealed. The left eye developed vitreous hemorrhage, retinal detachment, bullous keratopathy, and blindness, but the intraocular pressure is 20 mm Hg.

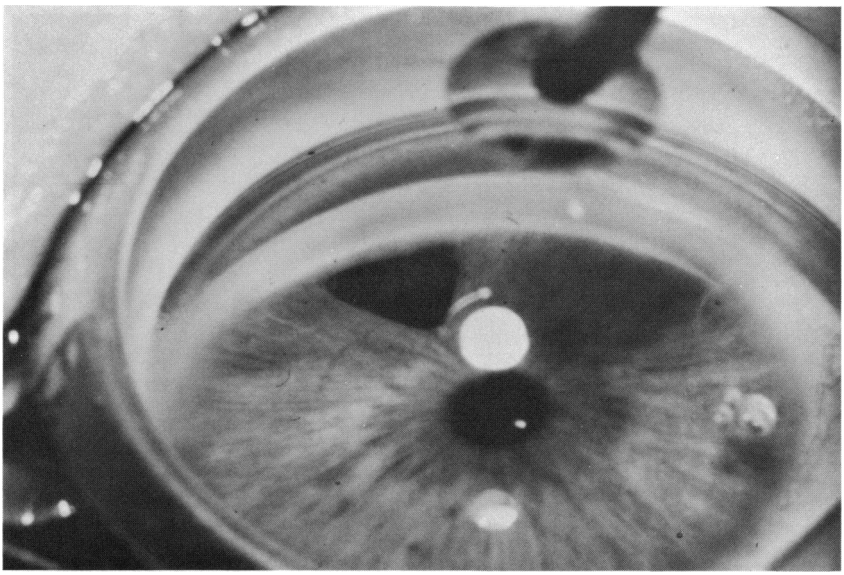


FIGURE 8

Case 6. Gonioscopic view of OD showing sealed angle and open peripheral iridectomy through which the lens equator appeared to touch the ciliary processes.

was that of chronic angle-closure with poor control on maximum medical therapy. Peripheral iridectomy was performed too late to control the glaucoma and the use of strong miotics probably let to the final acute angle-closure. The benefit from the lens extraction in the left eye was negated by the vitreous hemorrhage and exudative retinal detachment produced by the posterior sclerotomy. Lens extraction in the right eye was complicated by massive choroidal effusion and retinal detachment, and the prognosis at the present writing is still uncertain.

CASE 6

A 42-year-old plant worker had always been very farsighted but had fairly good vision until he presented with gradual painless impairment of vision in the left eye for eight months, and occasional halos around lights viewed by his right eye. Initial examination revealed the vision and glasses to be as follows: OD +12.50 sphere 20/60, OS +11.25 sphere +2.75 cyl axis 165° light perception. The eyes were not inflamed. The corneas were small, the anterior chambers very shallow and gonioscopy showed in the right eye a grade I angle which was 50% closed, and in the left eye closed 360°. Intraocular pressures were 32 in the right eye and 65 mm Hg in the left eye by applanation. Through miotic pupils the right disc was normal and the left disc was pale.

A peripheral iridectomy was performed on the right eye and three days later a peripheral iridectomy and posterior sclerotomy was performed on the left eye. The latter procedure was accompanied by a vitreous hemorrhage.

Intraocular pressure responses to the surgery were good for approximately two weeks but then became elevated again to 32 in the right eye and 23 mm Hg in the left eye. Gonioscopy in the right eye showed a closed angle except for a small area at the 5 o'clock position. The left eye developed bullous keratopathy and eight months later light perception was lost.

Five years after surgery was performed an examination showed the vision to be 20/100 in the right eye and no light perception in the left eye (Figure 7). Horizontal corneal diameters were 11.1 mm. Anterior chamber depth in the right eye was 2.30 mm and gonioscopy showed the angle to be closed by synechias 360°. In the surgical coloboma the lens equator seemed to be in contact with the ciliary processes; early nuclear cataract was present. There was marked bullous keratopathy in the left eye. With continued maximal medical therapy to the right eye, intraocular pressures were 28 in the right eye and 20 mm Hg in the left eye. The visual field in the right eye was normal.

COMMENT

A painless, progressive, chronic angle-closure resulted in marked damage to angle structures and vision at the time of first discovery. Posterior sclerotomy in the left eye resulted in vitreous hemorrhage. Peripheral iridectomy in the right eye probably deepened the anterior chamber, but was followed by the development of additional peripheral anterior synechias. The glaucoma in the right eye is no longer controlled with maximum medical treatment and the patient should have a filtering operation.

DISCUSSION

The eyes of the six patients in this series fit the definition of "nanophthalmos" in that they are smaller than normal in all dimensions. The eyes are extremely hyperopic with a congenital axial hypermetropia ranging from +11.00 sphere to +16.00 sphere. This was confirmed by ultrasonography in most of the patients. In early life the corrected vision was normal in all patients except in Case 1 (congenital exotropia and amblyopia ex anopsia of the left eye) and in cases 2 and 7 (probable macular hypoplasia). Although quantitative measurements varied from patient to patient the horizontal corneal diameters were all 11.1 mm or less and the anterior chamber depth measurements were 2.45 mm or less. The average normal corneal diameter in the adult is 11.8 mm,[11] and the average anterior chamber depth in hyperopes between the ages of 45 to 50 is 3.0 mm.[12] The relative large size of the lens resulted in a forward

TABLE III: CLINICAL CHARACTERISTICS OF NANOPHTHALMOS IN SIX PATIENTS (12 EYES)

Age	31-56 years (average 44.8) At onset of glaucoma (average 42.6)	
Vision prior to glaucoma	20/30 or better except in case 1 OS with amblyopia ex anopsia and 20/70 in both eyes of two patients (cases 2 and 6) with presumed macular hypoplasia	
Refraction	+11.00 sphere to +16.00 sphere	
Horizontal corneal diameter	10.5 mm to 11.1 mm	Normal adult age 40-50 11.8 mm
Anterior chamber depth	With glaucoma 1.50 mm to 2.30 mm Without glaucoma 2.30 mm to 2.45 mm	Normal hyperope age 40-50 ± 3.00 mm

position of the iris-lens diaphragm. On gonioscopy this produced a "vesuvio iris" appearance and made study of the angle very difficult. In the three eyes which did not have glaucoma (Cases 1 and 2) the angle was classified as "slit opening" in Case 1 and grade I in Case 2. In both patients the angle structures were seen only by tilting the lens and the trabecular meshwork and iris recess could be only partially visualized. No peripheral anterior synechias could be seen in these eyes and mydriatic provocative tests were negative. Unfortunately, careful gonioscopic studies on the remaining nine eyes were not performed until glaucoma was firmly established or treatment delayed.

A peculiar finding in all patients was the presence of a wide pulse pressure upon tonometry and tonography which made interpretation of the end point very uncertain. The pulsation of the Schiøtz pointer or the applanation half circles was present even when the intraocular pressure was low, and in the presence of high pressure would swing as much as 15 mm Hg on the scale. This pulse pressure was not associated with a wide systemic pulse pressure. It was always greater when the patient was emotionally upset or tense and was especially marked in Cases 4 and 5. The highest reading, which was associated with cardiac systole, was the one always used in determining the intraocular pressure.

The explanation of the tonometric ocular pulsation in these patients is not known but in the opinion of the author it is due to decreased ocular distensibility produced by the small volume of the globe, with its reduced aqueous volume and reduced choriodal vascular bed, which prevents dissipation of the cardiac systolic pulse. In some way this vascular abnormality must have something to do with the unexpected complications which follow anterior segment surgery, especially the occurrence of

TABLE IV: RESULTS OF PERIPHERAL IRIDECTOMY FOR GLAUCOMA IN NANOPHTHALMOS

Case	Glaucoma Benefitted	Clinical Course
#2 OS	No	Lens extraction necessary
#4 OD	Yes	Blind from posterior sclerotomy
#5 OS	No	Lens extraction necessary
#6 OD	No (?)	Secondary peripheral anterior synechia
#6 OS	Yes	Blind from posterior sclerotomy

TABLE V: RESULTS OF FILTERING OPERATION FOR GLAUCOMA IN NANOPHTHALMOS

Case	
#2 OS	Glaucoma not benefitted. Lens extraction necessary for control
#5 OD	Trabeculectomy combined with lens extraction followed by immediate massive choroidal and retinal detachment but control of glaucoma

retinal and vitreous hemorrhage after posterior sclerotomy in three patients (Cases 4, 5, and 6) and possibly with the tendency to choroidal effusion (Cases 3 and 5). In regard to the posterior sclerotomy, the surgeons were certain that the needle had not penetrated to the opposite wall of the eye. One must assume that the sclerotomies were performed too far posteriorly in these shortened eyes.

What type of glaucoma do these patients have, and is the nanophthalmos a contributing cause in its development? Typically, these patients develop a slow painless impairment of vision in one eye or are found to have an elevated intraocular pressure on a routine examination. Examination at this time usually reveals varying degrees of angle-closure and peripheral anterior synechias consistent with the pressure elevation. There will not be a completely open angle in the presence of an elevated pressure. Although therapeutic response to miotics and carbonic anhydrase inhibitors will seem to be effective for a short while, the glaucoma will soon become refractory and additional peripheral anterior synechias will appear. These responses alone point to the fact that we are dealing with a form of "creeping" angle-closure glaucoma and not chronic simple glaucoma with narrow angles.[13,14] The use of strong miotics make the condition worse, and in Cases 3 and 5, there occurred superimposed attacks of acute, painful, inflammatory angle-closure; no doubt due to the inflammatory reaction and pupillary block.

The dilemma facing the surgeon in such cases is produced by the fear of malignant glaucoma which might follow anterior segment surgery because of the shallow anterior chambers. This was the principal reason

TABLE VI: RESULTS OF LENS EXTRACTION FOR GLAUCOMA IN NANOPHTHALMOS

Case	Glaucoma Benefitted	Final Vision	Clinical Course
#2 OS	Yes	20/100	Two previous glaucoma operations
#3 OS	Yes	20/200	Late choroidal and retinal detachment
#3 OD	Yes	20/30	No further treatment
#5 OS	Yes	NLP	Blind from posterior sclerotomy hemorrhage
#5 OD	Yes	5/400	Immediate choroidal and retinal detachment

for deferring early peripheral iridectomy in several cases. The surgeon in Case 3 settled the problem by performing clear lens extraction and sector iridectomy in both eyes (seven years apart), and obtained complete control of the glaucoma in both eyes and normal vision in one eye. Five other eyes had peripheral iridectomies performed at a late stage with almost complete angle-closure. In two of these eyes (Cases 4 OD and 6 OS,) posterior sclerotomy was performed at the same time and although the glaucoma is no longer present, the eyes are blind from operative vitreous hemorrhage and complete exudative retinal detachment with vitreous bands. In Case 2, an unsuccessful peripheral iridectomy was followed in five months by an unsuccessful filtering operation which was followed in sixteen months by a lens extraction and glaucoma control. In Case 5, an unsuccessful peripheral iridectomy was followed in ten months by lens extraction and posterior sclerotomy which alleviated glaucoma but resulted in blindness from vitreous hemorrhage and exudative retinal detachment. In Case 6, peripheral iridectomy performed at a time when the angle was 50% closed succeeded in deepening the anterior chamber but five years later the glaucoma was still not controlled under maximum medical therapy and the angle was sealed 360°.

In none of the five eyes with peripheral iriedectomy did the dreaded acute postoperative malignant glaucoma develop. In retrospect, the two eyes which had posterior sclerotomy and vitreous hemorrhage might have fared better by risking simple peripheral iridectomy.

The possible role of a ciliolenticular-block mechanism in these cases is unclear. In Case 5, a periphery iridectomy failed to benefit the glaucoma but there appeared to be an adequate space between the lens equator and ciliary processes when viewed through the iris coloboma. In Case 6, with a similar failure of the iridectomy, the lens equator did seem to touch the ciliary processes. Unfortunately, the effect of cycloplegic drugs was not tried in either eye.

Filtering procedures were performed on only two eyes. In Case 5, a trabeculectomy was combined with lens extraction and has appeared to

function during the eventful postoperative choroidal effusion and retinal detachment. In Case 2, an anterior lip sclerectomy was performed five months after peripheral iridectomy but failed to control the glaucoma until lens extraction was performed sixteen months later.

In addition to the apparently hopeless exudative retinal detachments and vitreous bands which followed posterior sclerotomy in Cases 4 and 6 (peripheral iridectomy) and Case 5 (lens extraction), postoperative lens extraction choroidal effusion and nonrhegmatogenous retinal detachments occurred in two eyes (Case 2 and Case 5). The details of the management of these complications will not be stressed in this presentation but have been the subject of publications by Schepens and Brockhurst.[8,9,10]

Chiefly from the clinical manifestations and course the author believes that the glaucoma occurring in nanophthalmos represents an extreme in the spectrum of primary chronic angle-closure glaucoma and is not chronic simple glaucoma with narrow angles. The increased lens thickness and chamber shallowing that occurs with age is more apt to produce chronic angle-closure in the small, nanophthalmic eye than in the normal sized eye because of the relative large size of the lens. Despite the real danger of postoperative ciliary-block glaucoma, peripheral iridectomy should be beneficial in the early stages before damage has occurred to the outflow mechanism. The use of miotics probably worsens the condition by reducing pupillary flow of aqueous and possibly by encouraging ciliolenticular or ciliovitreal block,[15] but this cannot be definitely documented or proven nor has the effect of cycloplegic drugs been tried in any of the present cases. In the advanced stage of glaucoma, with a shallow anterior chamber and closed angle, filtering operations will probably not succeed and one must resort to lens extraction. In the present series, posterior sclerotomy combined with peripheral iridectomy (two cases) or lens extraction (one case) has resulted in vitreous hemorrhage and exudative retinal detachments and vitreous bands. Lens extraction has also been shown in two out of the five cases to be followed by choroidal detachment and effusion and by nonrhegmatogenous retinal detachment.

SUMMARY

Patients with nanophthalmos are prone to develop a chronic painless type of glaucoma in middle age, probably due to the natural increase in the size of the lens which is already relatively too large for the small eye. Although the underlying mechanism is obscure, a slowly progressive "creeping" chronic angle-closure is postulated, but gonioscopic evaluation

is difficult due to the shallow anterior chamber, with grade I and slit angles. Response to medical treatment is poor and miotics may even make the condition worse by producing relative pupillary block and by relaxing the lens zonule. Ordinary glaucoma surgery is to be avoided in nanophthalmos because of the fear of postoperative ciliary-block malignant glaucoma. Peripheral iridectomy performed in five eyes at an advanced stage of the chronic angle-closure did not facilitate glaucoma control in three eyes, and in two eyes in which the operation was combined with posterior sclerotomy, the eyes became blind from vitreous hemorrhage. Lens extraction in five eyes controlled the glaucoma but was followed by choroidal effusion and nonrhegmatogenous retinal detachments in two eyes and blindness in another eye when combined with a posterior sclerotomy.

No firm therapeutic recommendations can be made on the basis of the author's experience in the six reported cases. Conventional medical therapy seems ineffectual even when begun early in the glaucoma. Conventional glaucoma surgery must be performed early, before permanent damage to the outflow mechanism occurs but removal of the lens must be anticipated. The surgeon must also remain aware of the high incidence of serious posterior-segment complications which inexplicably follow glaucoma or lens surgery in nanophthalmos, as described by Brockhurst.[8]

ACKNOWLEDGEMENTS

The author wishes to express his thanks to Doctors O. B. Carlisle, Richard Brown, and Richard Connell for permission to include their cases, and especially to Dr. John C. Rieser for his advice and guidance in this study.

REFERENCES

1. Duke-Elder S: *Congenital Deformities*, in System of Ophthalmology, (St. Louis, C V Mosby Co, 1964,) vol III, Part 2, pp 488-495.
2. Wolff E: A Family With Microphthalmos. *Proc Roy Soc Med* 23:45-48, 1930.
3. Leyhdecker F: Eine Familie mit Mikrophthalmia Congenita. *Albrecht von Graefes Arch Klin Ophthalmol* 139:790, 1938.
4. Hatcher WF: Extreme Axial Hyperopia. *Arch Ophthalmol* 48:161-162, 1962.
5. Herman P: Le Syndrome microphthalmie-Retinite-Pigmentaire-Glaucome. *Arch Ophtalmol (Paris)* 18:17, 1958.
6. Vetter J: Ablatio Retinae als Zweiterkränkung bei Mikrophthalmus und Glaukom. *Klin Monatsbl Augenheilkd* 134:421-426, 1959.
7. O'Grady RE: Nanophthalmos. *Am J Ophthalmol* 71:1251-1253, 1971.
8. Brockhurst RJ: Nanophthalmos with Uveal Effusion: A New Clinical Entity. *Trans Am Ophthalmol Soc* 72:371-403, 1974.

9. Schepens CL, Brockhurst RJ: Uveal Effusion I, Clinical Picture. *Arch Ophthalmol* 70:189-201, 1963.
10. Brockhurst RJ, Lam KW: Uveal Effusion II, Case Report with Analysis of Subretinal Fluid. *Arch Ophthalmol* 90:399-401, 1973.
11. Wilmer HA, Scammon RE: Corneal Dimensions in Newborn and Adult. *Arch Ophthalmol* 43:599-620, 1950.
12. Calmettes L, et al: Etude de la Profundeur de la Chambre Anterieure. *Arch Ophtalmol (Paris)* 18:513-542, 1958.
13. Lowe RF: Primary Creeping Angle-Closure Glaucoma. *Br J Ophthalmol* 48:544, 1964.
14. Pollack IP: Chronic Angle-Closure Glaucoma. *Arch Ophthalmol* 85:676-689, 1971.
15. Wilkie J, Drance SM, Schulzer M: The Effects of Miotics on Anterior Chamber Depth. *Am J Ophthalmol* 68:78-83, 1969.

DISCUSSION

DR ROBERT N. SHAFFER. We are indebted to Dr Calhoun for bringing to our attention this rare and devastating type of angle-closure glaucoma. The hallmark of the condition is a small eye with a shallow anterior chamber, chronic angle-closure, and a marked hyperopia of +10.0 to +20.0 diopters. These eyes have a relative pupillary block and thus are helped by iridectomy. Some have a ciliary-block malignant type of glaucoma as shown by the continued high pressure and flat anterior chamber after filtering surgery. We have had two eyes like this whose anterior chambers could be kept formed and pressures under control only by the continuing use of topical atropine after iridectomies had been performed. It would seem logical to use atropine as a medical trial before resorting to surgery on these eyes which characteristically have such a catastrophic postoperative course. I would like to ask Dr Calhoun if atropine drops were used either preoperatively or postoperatively in his patients.

From a surgical point of view these eyes often respond like ciliary-block glaucoma, but then tend to suffer the unique complications of choroidal edema, intraocular hemorrhage, and retinal detachment. How can we explain these complications which result in blindness in half of the eyes operated upon?

I would like to suggest a hypothesis for the consideration of Dr Calhoun and also Dr Brockhurst whose thesis for this Society in 1974, was Nanophthalmos with Uveal Effusion. The hypothesis is that the surgical complications stem from choroidal engorgement which is due to impairment of venous drainage through the thickened scleras of these nanophthalmic eyes.

Dr Calhoun has described a peculiar pale mottled appearance of the fundus and a high pulse pressure in these eyes. This is easily seen in their tonograms, with swings in pressure as great as 15 mm Hg between systole and diastole. In Dr Brockhurst's case in which histopathologic studies became possible, the choroid was very thick and edematous and the blood vessels were engorged. There were diffuse infiltration of the choroid with mononuclear cells. The filling of this choroidal sponge in systole can produce the high pulse pressure just as it does with a choroidal hemangioma. The sclera was very thick, measuring 2 mm, which is similar to that noted in other cases at operation. It seems probable that it is this choroidal engorgement which leads to the permeability of choroidal

vessels, choroidal effusions, intraocular bleeding, and then retinal detachments which typically complicate the surgery of nanophthalmos.

I have been reminded of the choroidal edema with angle-closure which can result from encircling bands in retinal detachment surgery, as reported by Dr Ariah Schwartz at the 1974 Doheny meeting. These bands compress the vortex veins and result in choroidal effusion. Is it possible that the thickened sclera of these eyes impedes posterior flow of venous blood? Surgery on these eyes results in the choroidal effusion. If a posterior sclerotomy is performed, bleeding then occurs, leading to retinal detachment.

Whether this hypothesis is correct or not, these eyes do poorly surgically. Four of Dr Calhoun's eight cases; five of Brockhurst's 10 cases; and two of four eyes in our experiment ended with little or no vision. It would therefore seem wise to employ medical therapy as long as possible. Iridectomy followed by corticosteroids and topical atropine seems logical as an initial surgical procedure. The success rate of filtering surgery or lens removal is considerably below 50%.

It has been a privilege to discuss this interesting paper.

DR A. D. RUEDEMANN, JR. Mr President. Members and Guests. I would just like to ask two questions. Dr Calhoun alluded to the relative size of the lens in these cases. It has been my clinical impression that the lenses are at least normal size and appear larger in respect to the eye that you are attacking and secondly; I would ask if he used ultrasound in this regard. Finally, I would ask if there were any other congenital abnormalities associated with these cases. Thank you.

DR ROBERT J. BROCKHURST. Mr President. Secretary. Members and Guests. I, like Doctor Shaffer, was impressed with this morning's program to hear two papers which involved choroidal detachment. Choroidal detachment has been a subject which has interested me for sometime, and I've been surprised that in the literature very little is actually devoted to this problem. In general, it is usually secondary to hypotony; complicating uveitis, trauma, or surgery. Doctor Garrity and Doctor Harbison's paper indicates that an increase of pressure in the venous drainage system creates a relative hypotony which is another mechanism. I believe Doctor Shaffer is quite right that the thick sclera which is seen in patients with nanophthalmos may become more rigid during life increasing the relative resistance to venous drainage. This would encourage the development of a peripheral choroidal detachment or effusion which would further narrow the already compromised angle in these patients. In addition to the points mentioned this morning, the choroidal detachment in uveal effusion may be interpreted as a tumor. I have observed one patient in whom a melanoma was diagnosed and the eye was enucleated. Second, the secondary retinal detachment which occurs is absolutely untreatable by any known surgical method. Even drainage of the subretinal fluid, performed very carefully and completely, will be followed within 24-48 hours by a total detachment such as shown in the photograph. Finally, I would like to thank Doctor Calhoun very much for allowing me the opportunity to see his paper.

DR DAVID SHOCH. Mr President. Ladies and gentlemen. I enjoyed listening to this paper. In 1971, Dr Richard O'Grady of our Department described the pathology of two eyes from a patient with nanophthalmos (*Am J Ophthalmol* 71:1251, 1971). His point, to answer Dr Ruedemann's question, was made in that paper in that the corneas measured some 9 mm in diameter and the lenses were, in fact, of normal size. It was our feeling that this was the cause of the problem. It is noteworthy that these eyes had several glaucoma procedures, all of which had failed and resulted in bilateral enucleations. It was our feeling that this represented the obverse, in a sense, of anterior megaophthalmos where you have a very large cornea with perhaps a normal lens. Here we have what might be termed anterior microphthalmus where there is a small anterior segment but a normal lens. It is this disproportion that is fundamentally the cause of the glaucoma; therefore, the appropriate treatment for the glaucoma of nanophthalmos is lens extraction.

MR JOHN DOBREE. Mr President. Members. First of all, may I say what a tremendous privilege it is to be here and what a wonderful meeting this is. Now in discussing this , firstly may I say that I have never seen this complication; but I think I would just offer a suggestion as to a possible surgical procedure. I think that this situation might very well respond to a trabeculotomy. Now this is one of the least traumatizing anti-glaucoma procedures. Sometimes it is impossible to do because one cannot find the canal. However, there is a minimal amount of bleeding, a minimal amount of trauma, and you can do it very easily, for example, in buphthalmos and in peripheral anterior synechiae where you want to open the angle. I would have thought, sir, that this would be a possible procedure in this case, and perhaps one of the previous speakers would like to comment on that. Thank you.

DR ROBERT MOSES. Mr President, Mr Secretary. Just a very brief comment on a possible explanation for the very large pulse pressures noted. The tension within the wall of a cylindrical or spherical vessel, and I mean the actual stretching force on the containing wall of the vessel, is proportional to the radius of the vessel and inversely proportional to the wall thickness. That is, the smaller the eye the less actual tension will be exerted on the sclera-cornea by a given pressure. Consider for a moment that in industry, gases under high pressure are put in small radius cylinders, since there is less tension induced in the walls by the high pressure. In the case of an eye, a change in intraocular pressure as the 30 mm or 40 mm of pulse pressure would be withstood by a small eye much better than a large one; the small eye would expand less with the increase in intraocular pressure with the pulse and the fact that the sclera-cornea distends less would damp the pulse pressure less in a small eye than in a large one and might help explain some of the large pulse pressure. This explanation assumes that the sclera-cornea of the small eye is as thick as that of the larger one.

DR F. PHINIZY CALHOUN. I deeply appreciate the interest shown by the discussers. First of all, I would like to answer Doctor Shaffer and Doctor Brockhurst together regarding the scleral thickness. In only one patient, was the scleral thickness actually measured. It was not noted or measured in the patients who had posterior sclerotomy because it was not thought of at that time. In other words, scleral thickening was noted only in case 5 after lens removal and I don't think that in any case it has been found to be present before glaucoma surgery or lens surgery has been performed. In other words, we feel that it is postoperative swelling of the sclera associated with the choroidal effusion and not a pre-existing thickening of the sclera; but this certainly has not been proven. Unfortunately, none of these patients have either been treated preoperatively with atropine. The role of a ciliary block mechanism in these cases and the effect of atropine or proper therapy was really not tried in any of our cases.

Doctor Shaffer, I am sure that there must be some vascular abnormality somewhere in the choroid, or elsewhere. I am sure that is your suggestion about the possible cause. Doctor Ruedemann, the lens size in the one case (the next to the last case) was measured immediately after removing it from the eye and it was slightly smaller. The lens is usually considered normal in these cases but in this particular case it was only relatively normal since the dimensions were 8.5×8.5×4 rather than closer to 9 mm in diameter. Usually the lens, of course, is much more normal size than the rest of the eye. Ultrasound was used in most of the patients for determining the AP measurements and it was very helpful.

The reason these cases were called nanophthalmos is because there is no other known ocular anomaly associated with it. In other words, this is supposed to be an otherwise normal eye. Doctor Shoch mentioned O'Grady's case. There were two eyes that were removed and pathologic examination did not show thickened sclera. No lens removal had been performed in either one of those eyes.

Mr Dobree, I appreciate your comment regarding trabeculotomy. This has not been thought of or tried, and I think we have to consider this as one of the possibilities in performing anterior segment glaucoma surgery. The fear is that anything that will open the eye or reduce the pressure will further endanger the narrow slit-like angle that exists. One of the reasons surgery of any kind is delayed is because of the fear of producing angle-closure or malignant glaucoma following the surgery.

Doctor Moses, I appreciate your comments about the explanation for the wide pulse pressure which agrees with my own ideas; but am unable to find a true explanation according to a known law of physics or physiology.