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HYDROPTHALMOS—A BIBLIOGRAPHIC, CLINICAL AND PATHOLOGIC STUDY.

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HYDROPTHALMOS is a more or less uniform increase of all the diameters of the infantile eyeball, due to the expansion and consequent thinning of the elastic sclera under increased intraocular pressure. It occurs at birth or in early infancy, but cases of later development have been reported. It may be unilateral or bilateral.

Considerable confusion has arisen from the multiplicity of names used to denote this condition. We find in literature all of the following titles: Hydrophthalmos, hydrophthalmos congenitus, hydrops oculi, dropsy of the eye, bupthalmos, "ox-eye," megalophthalmos, megalocornea, keratoglobus, cornea globosa, globose cornea, glaucoma congenitum, congenital glaucoma, infantile glaucoma and infantile secondary glaucoma.

With such a variety of titles, the difficulties of preparing a bibliography and collecting the various expressions of opinion relative to this subject may be readily understood. Unfortunately no author has called attention to this chaotic state of the nomenclature; it is only by securing the best descriptive name and uniformly adhering to it that universal bibliographic research may be systematically pursued.

Hydrophthalmos is undoubtedly the best name to use. It describes a general underlying condition without refuting any theory of causation or misinterpreting any local expression of the disease. Hydrophthalmos congenitus and hydrops oculi are not of such common usage and consist of two words.

Dropsy of the eye is too colloquial. Buphthalmos and its English equivalent "ox-eye" have been extensively employed, but they are not scientific, and, by many lexicographers, are used only to express that condition in which the eyeball is so distended as to prevent closure of the lids. Megalophthalmos simply means large eye. Megalocornea, keratoglobus, cornea globosa and their English equivalent, globose cornea, refer only to the state of the cornea. In view of the difference from glaucoma in pathologic anatomy, and the denial of the etiologic similarity by some observers, glaucoma congenitum, congenital glaucoma, infantile glaucoma and infantile secondary glaucoma will always excite antagonism.

Hydrophthalmos means increase of the fluid contents of the eyeball, irrespective of the diverse consequences thereof. It would be better perhaps to use a word indicating uniform expansion of the ocular tunics, but there is no such word in current use, and hydrophthalmos has the prestige of being the original term, the one most commonly used at present, and, in addition, one that permits of infinite subdivision.

HISTORY.—Hydrophthalmos has been recognized as a distinct affection for many years. There are numerous references to it in the medical literature of the seventeenth and eighteenth centuries, as a glance at the bibliography will show. However, it is very likely that some of the older cases reported as hydrophthalmos were really instances of large anterior staphyloma, exophthalmos, intraocular tumor, etc.

Borellus⁵ mentioned hydrophthalmos, and advised treatment by blistering the nape of the neck and using a collyrium of white vitriol. Beyer,⁴ in 1774, reported a curious case, and in 1775, Grygerus¹⁹ discussed the subject. In 1776, Terras³⁵ called attention to the value of paracentesis in the treatment, and Mauchart,²⁷ Marchan,²⁶ Heister,²⁰ and an author in the *Ephemerides*,¹¹ made similar observations. In the *Acta Naturæ Curiosorum*, Vol. VIII, obs. 44, there is mention of a recurrent case (likely secondary glaucoma). Brandis⁶ reported the occurrence of the disease in scrofulous patients. Cheston⁹ described a fatal case with caries of the orbit. Gendron¹⁷ men-

tioned the enlargement of the eyeball to the size of a hen's egg. Over a century ago, Conradi¹⁰ attributed the disease to an accumulation of vitreous humor. Morand²⁹ wrote of a case in which "the tunica of the optic nerve was dilated and elongated." Observing the dryness and opacity of the cornea in this condition, Pellier³⁰ ascribed the cause to "cornea siccatus," and advised the use of emollients. Heister²⁰ dwelt upon the advantages of compression. Scarpa³² and Langlebert²² mentioned extirpation of the eyeball in hydrophthalmos, and in 1790, Langlebert²² made a similar observation. In 1781, Ford¹⁵ published the history of a case successfully treated, and in 1784, another¹⁶ in which the eye, after vision had been ruined, was reduced to the normal size by means of a seton.

Early in the nineteenth century, Luke²⁵ published a dissertation on hydrops oculi, and in 1806, Joli³⁷ issued a similar monograph. In the next few years, Beer,² Benedict,³ Rowley,³¹ and Thilow³⁶ described hydrophthalmos. In 1822, both Sturm³⁹ and Reeder issued papers upon the subject. In 1830, Bachman⁴³ published an extensive article on the causes, varieties and treatment of the condition. In 1832, Von Ammon⁴⁵ discussed the pathologic histology. In 1834, Masche,⁴⁶ Radecke,⁴⁷ and Gerson⁴⁸ appeared with papers; in 1836, Grellois;⁴⁹ and in 1841, Magdalener.⁵¹ In 1843, Flarer⁵³ brought forth a paper on "Buphtalmia." In 1853, the renowned Nélaton⁵⁹ delivered a clinical lecture upon this subject; Arlt⁶¹ devoted his attention to it in 1857. Articles were published in 1871 by Schirmer;⁶⁸ in 1876, by Raab;⁶⁹ in 1879, by Fano;⁷⁴ and in 1881, by Bergmeister.⁷⁶

In 1885, Grahmer⁸² contributed a valuable paper on the pathologic anatomy of the affection. In the following year, Gallenga⁸⁴ collected a number of cases. In 1892, Cross⁸⁸ issued a valuable statistical paper. In the same year, Kalt⁸⁹ studied the pathology. In the Hunterian Lecture, London, 1894, Treacher Collins⁹⁵ advocated the theory of congenital defect of development. In 1895, Snellen⁹⁶ discussed the treatment of conditions leading to hydrophthalmos. In the same year, Lodato⁹⁸ and Bockel⁹⁷ offered general contributions, and Angelucci⁹⁹ advanced the vaso-

motor theory. In 1896, Cabannes¹⁰⁰ and Bergmeister¹⁰¹ reported cases. In 1897, Geos,¹⁰³ a medical graduate of Paris, took the subject for his thesis. In 1898, Johnson¹⁰⁶ detailed the history of a most interesting series of cases occurring in the same family.

SYMPTOMS.—Hydrophthalmos generally appears in early infancy. The most prominent characteristic is the uniformly enlarged and protruding eyeball (more noticeable in unilateral cases by comparison). The lids seem insufficient and appear small in comparison to the ball; complete closure is often impossible, and part of the cornea may be exposed during sleep (lagophthalmos). Rotation of the globe is impeded, and all ocular movements are slow and incomplete.

The sclera is thinned by overstretching, and presents a peculiar bluish hue due to the uveal pigment showing through it.

The cornea is noticeably enlarged in all its diameters, and is of increased curvature, partially causing the extra protrusion. It is more or less insensitive. Very soon in the course of the disease, the cornea becomes hazy, progressing to varying degrees of cloudiness, although cases are on record in which the cornea has remained translucent (*keratoglobus pellucidus*).

The anterior chamber is generally deep, quite the reverse of the condition in adult glaucoma. However, in cases due to congenital irido-keratitis, the anterior chamber may become almost obliterated, as in adult secondary glaucoma. One of the cases pathologically studied by myself showed such a condition.

Unless in the process of active inflammation, the iris is generally more or less atrophied and, as a consequence of this and increased tension, the pupil is partially dilated and sluggish in reaction. If the lens is displaced or the zonule of Zinn is atrophied, the iris will be tremulous. Such a condition is also associated with a noticeably wabbling lens.

The media are usually too cloudy to permit of satisfactory ophthalmoscopic examination, but if the process is of long duration, it may be assumed that the papilla is excavated and the optic nerve atrophic.

There is considerable increase of tension, often

accompanied by severe pain. The infant seems irritable and may continually rub and scratch the eye and give other signs of local irritation. When it becomes older, it often complains of a dull pain in the ball.

CLINICAL COURSE.—The condition is recognized at birth or shortly after, and the incipient stages are usually intrauterine. There is a slow but progressive enlargement of the eyeball in all its diameters, with corresponding diminution in vision, due to opacity of the cornea, disorganization of the vitreous, detachment and stretching of the retina, and excavation of the nerve-head.

Sometimes, after a certain stage in the course has been reached, the progress of the disease becomes very slow or entirely checked, and in such cases the tension remains stationary or gradually diminishes to slightly above normal. The enlargement of the eyeball ceases, but in the absence of operative interference, the globe seldom becomes much smaller. The degree of preservation of vision depends upon the state of the cornea and vitreous and the amount of injury to the retina and optic nerve.

Unfortunately, in the majority of cases, the tendency is to progressive enlargement, the cornea becomes more and more opaque and may perforate or entirely slough; the optic nerve becomes completely destroyed, and the interior of the eye disorganized, causing total blindness. In rural districts, where medical attention is not common and ocular surgery is in a primitive condition, a case of hydrophthalmos is sometimes allowed to attain enormous proportions. In the classical case of Gendron,¹⁷ already cited, the eye was stated to be "extensus ad magnitudinem ovi gallinacei." This case was likely one of neglected large scleral ectasiæ with corneal staphyloma, and not true hydrophthalmos.

ETIOLOGY.—The nature of this disease has always been a matter of dispute. The direct cause of the physical changes is increased intraocular pressure. In the child, the sclera is very elastic, and under increased tension stretches uniformly; in the adult the sclera is tough and resistant and, unless weakened by

inflammation, it permits of expansion only at its weakest point—the lamina cribrosa (and possibly at the entrances of the venæ vorticosæ and anterior ciliary vessels). It is for this reason that glaucoma of the adult differs in its physical and pathologic appearances from increased intraocular pressure in children.

From its clinical course, the incipient stages of hydrophthalmos must be intrauterine. Dehenne⁸⁰ maintains that infantile glaucoma or hydrophthalmos may also occur in adults if the conditions are the same as those in a child, viz: if the sclera is elastic and yields in its entire surface. Gallenga⁸⁴ reports 3 such cases of acquired hydrophthalmos in his clinic at Turin. Dehenne⁸⁰ adds that on the other hand, when the sclera and cornea of a child have been extended ad maximum, excavation of the optic nerve begins—and a condition similar to the glaucoma of adults develops.

The two possible direct causes of increased intraocular tension are: first, increased secretion; and, second, obstruction to excretion. Increased secretion may arise from inflammatory changes in the region of the ciliary body, or from general vasomotor and circulatory disturbances. Obstruction to excretion may be caused by partial closure of the iridic angle at the periphery of the anterior chamber or obliteration of the neighboring infiltration channels. Either of these conditions may be the result of an intrauterine inflammation, such as irido-keratitis; or may be noninflammatory, *i.e.*, the result of congenital malformation, such as failure of the iris to completely separate from the cornea or of the canal of Schlemm to become completely patulous.

Every one of the possible causes has had its adherents and advocates, sometimes to the exclusion of all others, and a veritable war of argument has been waged. Doubtless each reporter has been in a measure correct, and the cases may be divided according to the condition or combination of conditions causing them. The different observers have, as a rule, based their conclusions on the particular case or cases examined, but all of these cases are not similar, and

there is no universally uniform cause. The chief symptoms are the result of intraocular pressure, but the exciting cause thereof differs in the individual cases. Therefore the general term hydrophthalmos is better than a term referable to a special cause or a special consequence.

In order to present a full discussion of the etiology of hydrophthalmos, the different theories and the opinions of their prominent advocates are cited below.

Predisposing Causes.—In the discussion of hydrophthalmos, various predisposing causes have been advanced. There is abundant evidence that *heredity* plays a prominent part in many cases, although it is sometimes difficult to substantiate the suspicion. W. B. Johnson,¹⁰⁶ of Paterson, N. J., reports the development of 3 cases of the disease in the same family, a circumstance certainly suggestive of the existence of some underlying family tendency for the production of this condition. This, however, could not be practically demonstrated, and, strange to say, the first 3 children were perfectly healthy.

Syphilis has long been suspected in the etiology of this disease. As long ago as the last century Brandis⁶ spoke of the "scrofulous" origin of the disease, and recent observers have seen the condition in children who exhibited all the associate signs of congenital syphilis. Interstitial keratitis being such a constant symptom of congenital syphilis, there is no doubt that some confusion has arisen, but authentic cases have been recorded, and I have definite knowledge of a case of hydrophthalmos in a child of 14 months with genuine interstitial keratitis. It is in the inflammatory type of the disease that we would suspect hereditary syphilis as a cause.

Increased Secretion by Inflammatory Changes.—According to some observers, and particularly Kalt,⁸⁹ who made extensive anatomic investigations on this subject, hydrophthalmos is a sequence of a chronic irido-choroiditis (uveitis) which leads to a gradual obliteration of the vessels of the uveal tract. This causes obstruction of the largest portion of the choroidal capillaries and increases the pressure in the ciliary arteries, from which arises the increased transudation of fluid.

Increased Secretion and Vasomotor Disturbance.—Angelucci⁹⁹ describes a case in a child of 6 years with tachycardia, frequent intermitting of the heart-pulsation, and congenital hydrophthalmos in both eyes. These cardiac disturbances have been observed by him in all his cases of congenital hydrophthalmos; with sometimes, also, valvular insufficiency and dilation of the left heart, arterial hypertension, and subjective and objective sensations of heat on the head and face. In these cases the character is always emotional and high-tempered. These facts lead the author to believe that hydrophthalmos is the result of vasomotor disturbances, and still more so, as trophic affections are observed in such eyes. It might be added that the pain in itself is sufficient to produce an irritable disposition in the child, and that the so-called trophic disturbances are readily explained by the increase of intraocular tension.

Similarity to Glaucoma.—Hydrophthalmos is regarded by many authors as a secondary infantile glaucoma, and some consider it a form of primary glaucoma occurring in children. Grahamer⁸¹ cannot agree with Horner, Mauther and other early observers in assuming that every case of congenital hydrophthalmos is one of congenital glaucoma. His investigations demonstrated the absence of inflammation and occlusion of the iris-angle—the test for the existence of glaucoma. He, therefore, believes that either an uveitis or a serous cyclitis is the primary cause, to which increased intraocular tension and other intraocular symptoms are secondary. The decrease in the caliber of the veins of Leber's plexus and obstruction of the spaces of Fontana further serve to augment the intraocular tension. In one of the cases pathologically studied by myself, observations similar to those of Grahamer were made.

Intrauterine Inflammation or Congenital Development.—Many authors believe that the disease is the result of an intrauterine irido-keratitis, causing the base of the iris to adhere to the cornea and closing the iridic angle. By some the presence of a low-grade keratitis and the atrophic condition of the iris are considered evidence for this belief. Again, there

is a class of modern observers who believe that the corneal changes are the result of trophic disturbances secondary to increased intraocular tension or of exposure by insufficient closure of the lids, and who attribute the general ocular condition to congenital defect of development. There is also evidence, already mentioned, to show that the corneal condition may be due in great part to interstitial keratitis of syphilitic origin.

In the Hunterian lecture, London, December, 1894, Treacher Collins⁹⁵ said that he believed that the peripheral adhesion of the iris and cornea was most likely a fault of development—failure in the separation in these structures, rather than a product of disease. In eyeballs affected with hydrophthalmos, though the angle is closed, the rest of the anterior chamber is deep, as fluid accumulates in the aqueous rather than in the vitreous. This shows that the primary obstruction is at the outlet of the aqueous chamber. In the case clinically studied in the present paper, signs indicative of present or past inflammation were absent, and, in addition, there were marked associate physical anomalies. In one of the cases pathologically studied, there was absence of the canal of Schlemm, presumably congenital, as there was not sufficient evidence of previous inflammation to cause the closure. However, before deciding upon congenital absence, it must be remembered that the canal of Schlemm and the spaces of Fontana are often obstructed by inflammatory conditions in the neighboring structures, and this is particularly so in infantile eyeballs.

PATHOLOGIC ANATOMY.—The increase in the size of the eyeball is due to the expansion of the elastic membranes of the infantile eyeball on their inner surfaces. Manz examined a case of congenital hydrophthalmos in a relatively early stage of development. He found both the anterior and posterior portions of the globe much enlarged, and the vitreous chamber partly filled with aqueous fluid. I have made somewhat similar observations.

The *cornea* is always more or less opaque, which, as already has been shown, is by some attributed to an

irido-keratitis in utero, and by others to trophic disturbances secondary to the increased intraocular tension. In other cases the corneal disease may be in a measure due to exposure, the globe being so distended as to prevent complete closure of the lids (*lagophthalmos*).

The *anterior chamber* is deepened in true hydrophthalmos instead of being shallow, as in glaucoma. There is abundant clinical and microscopic evidence to this effect, and I have definitely confirmed it both macroscopically and microscopically. However, in extensive inflammatory cases, the anterior chamber may be almost obliterated, as seen in one of the pathologic studies presented.

The *sclera* is much distended and thinned, allowing the uveal pigment to show through, causing the peculiar bluish appearance so characteristic of the disease.

The *choroid* and *retina* are stretched and tenuous, and may be detached and partially disorganized.

The *nerve-head* may be intact or may become excavated very soon in the course of the disease; this excavation progresses with the enlargement of the ball and increase of tension, until the nerve becomes entirely atrophic. Manz⁸³ could not find any excavation of the nerve-head in a case microscopically examined by him, and in one of the cases studied by myself, there was stretching, but no excavation of the nerve-head.

The *ciliary body* is swollen and the contained veins are markedly turgid. The *ciliary processes* become squeezed against the sclera and push the *iris* to the cornea and sclera, where it may remain adherent, and finally, with the ciliary body, atrophy, sometimes disappearing. Cabannes¹⁰⁰ showed a patient with hydrophthalmos and complete blindness. The right eye had been lost for a long time, the left eye was very prominent, extremely large, amblyopic, and the iris was absent. Lagrange explained the *aniridia* by atrophy of the iris, which was first compressed against the posterior surface of the cornea, and had then undergone sclerosing alterations and disappeared little by little as a distinct organ. It might be added

that it is possible that the aniridia was the result of defective development.

The *lens* retains its normal dimensions or diminishes in size. The pressure causing the sclera to expand is internal. The pressure upon the lens is exerted on its external surface. As the lens does not enlarge with the rest of the globe, the space between it and the margin of the ciliary processes becomes greater and greater and the *zonule of Zinn* is put more and more upon the stretch. If the general ocular enlargement is very great, the zonule is so stretched and thinned that it atrophies, and allows the lens to become unsteady or entirely luxated. This is evidenced in advanced cases by a trembling iris and wabbling lens. Dislocation of the lens is followed by its absorption in the vitreous and the usual disastrous consequences.

The *vitreous* is usually more liquid in consistency and full of opacities, the *aqueous* becomes more albuminous.

DIAGNOSIS.—Hydrophthalmos is readily differentiated. Its distinguishing marks are uniformly enlarged eyeball with insufficient lids in infantile life, the peculiar bluish sclera, enlarged and protruding cornea, the alteration in corneal transparency varying from a bluish, milky haze to complete opacity, increase of tension, atrophic or wabbling iris, tremulous or dislocated lens, and deep anterior chamber.

In keratoglobus or keratectasia, the enlargement is not uniform, and the cornea, and, possibly, the neighboring sclera are the parts involved. There is not always rise of tension, and in most cases a previous history of disease or injury is obtainable. Again, such cases would not likely date from birth. In cases of large scleral ectasia with corneal participation, the diagnosis might be confusing, but the irregular form of the staphylomata, and their history should readily distinguish them. Total scleral ectasia may occur at the same time with staphyloma of the cornea, but again, the irregular expansion and protrusion are diagnostic and there is history of previous injury or disease.

In keratoconus, the conical appearance is typical

and the cornea alone is involved. There is not usually marked increase of tension, and the cornea generally remains clear.

In glaucoma, the eyeball is hard but not noticeably enlarged, and the anterior chamber is shallow. The sclera has not the peculiar bluish appearance and there is not so much corneal involvement. Again, glaucoma is a disease of adult life, and the history of its progress is entirely different.

In exophthalmos there is no enlargement of the eyeball, simply protrusion. If the protrusion is the result of exophthalmic goiter, there are the associate circulatory symptoms, the enlarged thyroid and the inability of the upper lid to follow downward movements of the eyeball; if the result of orbital growths, the tumor may be defined behind the protruding ball. Again, in exophthalmos, vision is not necessarily involved, unless the optic nerve is pressed upon or stretched or the cornea becomes affected. Intraocular growths might be a source of confusion, but in childhood, glioma of the retina is really the only tumor likely to cause a mistake in diagnosis, and the peculiar ophthalmic appearance ("cat's eye") and the clinical history of this affection are proof conclusive.

PROGNOSIS.—The prognosis of hydrophthalmos is bad. Vision is almost invariably diminished to a considerable extent, although there are rare exceptions to this rule. Warlomont⁹⁴ reported a case of hydrophthalmos in a subject of 13 years with preservation of good vision. If the course of the disease is slow, or checked spontaneously or by operation, the amount of vision retained may be possibly preserved until the end of life; but, unfortunately, unless an operation is done in early infancy the cornea is so obscured and the retina and optic nerve so damaged that useful vision is gone. If the progressive enlargement of the eye cannot be checked, total blindness will ensure and, in addition, enucleation may be necessary on account of constant pain, excessive size, perforation, infection, or disorganization of the globe. It may be added that the prognosis is worse in the congenital form of the disease in which there is absence of the ordinary filtration channels.

SEQUELS.—The serious consequences of this disease are opacity, perforation and sloughing of the cornea, disorganization of the vitreous, atrophy of the iris and the ciliary body, opacity and dislocation of the lens, excavation of the nerve-head and consequent atrophy, intraocular hemorrhage, detachment of the retina and choroid, and, finally, disorganization of the whole ball with panophthalmitis, demanding immediate enucleation. In addition, the ocular muscles and the lids may become atrophic and partially paralyzed from want of proper use.

TREATMENT.—The majority of textbooks mention operative interference only to condemn it. Miotics are universally recommended without promise of much result. Minor operations, like repeated paracenteses, are not forbidden. In cases in which the increase of intraocular tension is dependent on synechiæ, iridectomy, before the iritic adhesions are too firmly formed, is recommended as a speedy means of relief. However, a search of the literature shows, among individual reporters, a far more favorable aspect for operative measures. But it must be remembered that it is usually the cases of fortunate issue that find their way into print.

In considering the operative treatment of hydrophthalmos, the pathologic anatomy of the distended eyeball must be borne in mind. The tunics are thinned and distended and the suspensory ligament (zonule of Zinn) is stretched, or, perhaps, ruptured. Again, there is great difficulty in operating upon infants. Hence, sclerotomy, and still more so iridectomy, is dangerous. There are the perilous possibilities of loss of vitreous (sometimes immediately after corneal section) and collapse of the globe, intraocular hemorrhage, and purulent infection of the vitreous. It is universally admitted that iridectomy and sclerotomy, done in the late stages of hydrophthalmos, are liable to be complicated with hemorrhage, and a future state of chronic and painful irritation.

With the italicized remark that *operations, to be of value, must be done in early infancy*, I forbear further comment, and proceed to present the views of individual reporters for and against operations.

Paracentesis is considered a plausible procedure by nearly every authority. Even in the last century Mauchart,²⁷ Heister,²⁰ Marchan,²⁶ and Terras³⁵ mentioned the good effects of single and repeated paracenteses. Juler⁹⁰ has performed paracentesis with improvement in a case in which he had been compelled to enucleate one eye on account of corneal perforation.

Seton, Injection of the Vitreous, etc.—In 1784, Ford¹⁶ successfully reduced the size of a sightless and drop-sical eye by means of a *seton*. In 1843, Flarer⁵³ tried this mode of treatment with good result. Masselon⁸⁵ showed, in Paris, a child, 22 months old, into whose eye de Wecker had introduced a small golden cannula, 20 months before, on account of increasing hydrophthalmos. The cannula was borne well; the cornea, which had been dull, cleared up; and, at the time of exhibition, tension was normal.

In 1855, Chavanne⁶⁰ *injected iodine* into the globe with little result.

Curious are the remedies of some of the older physicians. Heister used compression. Borellus⁵ mentioned blistering the nape of the neck and the use of a collyrium of white vitriol. In 1790, Langlebert²² reported a case treated by combined operation and cauterization.

Iridectomy and Sclerotomy.—Moralt, in 1869, reported a case of hydrophthalmos in which he had performed iridectomy, and, although not aware of any similar cases on record, he admitted the bad prognosis. Lodato⁹⁸ has made extensive clinical observations to prove the uselessness of iridectomy in the treatment of congenital hydrophthalmos. In common with other textbook authors, who are always wisely conservative, Schmidt,⁷³ in 1877, in his article in the Graefe-Saemisch Handbook, says: "In the secondary glaucoma which complicates cornea globosa (congenital hydrophthalmos), iridectomy is very dangerous and offers a poor prognosis." He adds that when, in these cases, rapid increase of intraocular pressure renders some kind of action necessary, it is best to try the effect of repeated paracenteses, which are certainly free from danger.

On the other hand, Dufour⁷⁷ advocates iridectomy. Gorecki⁸⁷ and Meyer perform it, and, with the present knowledge of the condition, do not consider it nearly so dangerous as formerly. Mellinger⁸⁶ reported two cases of iridectomy in hydrophthalmos with good results, occurring in children of 2 and 5 years respectively. In both cases he operated upon the left eye under chloroform and cocain. A broad coloboma was obtained; the diameters of the cornea decreased perceptibly after the operation. Derby⁷⁸ reported the performance of iridectomy in 3 cases of hydrophthalmos. In the first case, the disease had lasted 14 years and there was distinct optic atrophy at the time of operation. In the second case, the disease had lasted 10 years. In both cases, the beneficial effect of iridectomy was indisputable. In the third case, the operation was performed in an early stage of the disease, and the process was at once stayed. Eleven years had elapsed since the operation, and no further progress of the disease was manifest. Gorecki⁸⁷ presented, at a meeting of the Ophthalmological Society of Paris, a child of 3 upon whom he had performed iridectomy at the age of 10 months, and again at 14 months, with good result.

Bergmeister¹⁰¹ exhibited a boy of 13, upon whom he had done a successful iridectomy at the age of 6 months for "infantile glaucoma" (increased tension, diffuse opacity of the cornea, moderate enlargement of the ball, etc.). At the time of exhibition there was a normal anterior chamber, no excavation of the disc, and ability to count fingers nearby. The patient had had strabismus since infancy. Worthy of note in this case was an opacity of the equator of the lens in the coloboma, which Bergmeister believed to be due to the contact of the infantile lens with the operation-wound.

Fuchs⁹¹ mentions hydrophthalmos as one of the conditions indicating sclerotomy rather than iridectomy. In his graduation-thesis at Paris, Geos¹⁰³ says the treatment of hydrophthalmos consists in early operative interference—sclerotomy or iridectomy; the longer the delay, the smaller the chance of success. He is even enthusiastic enough over early operations

to say that hydrophthalmos may be cured by sclerotomy, whereas, for glaucoma this operation is only palliative. He adds that iridectomy is dangerous in eyes very much stretched or greatly enlarged, while it is useful in eyes that are very little distended in the early stages of the disease, in which cases it should certainly be tried if sclerotomy has failed. Geos also mentions that when an eye has become greatly enlarged it is better to do repeated sclerotomies than to attempt iridectomy; the larger the eye, the smaller ought to be the incision into the globe. Geos says that medicinal treatment by iodids, mercurials, miotics, etc., is useless, offers no chance of cure, and wastes valuable time. Miotics, however, are useful adjuncts to operations and should not be neglected.

W. B. Johnson¹⁰⁶ has reported a most interesting series of cases in the same family, in which the results attained by early operation were most gratifying and emphasized the necessity of immediate operative interference. The father and mother of Johnson's patients were healthy, and no significant family-history was obtainable. The father had a cataractous lens in his left eye. The first 3 children were perfectly healthy and had normal eyes. The fourth child was born with hydrophthalmos and was operated upon in Naples by de Vincentiis. A sclerotomy was performed at the second month. The fifth child was similarly operated upon at the second month with good result. In the meantime the family emigrated to Paterson, N. J., where the sixth child was born. She was a well-developed girl and was first examined in November, 1897, the parents believing that she was developing the condition which had appeared in her older sisters. At that time there was apparently but little advance of the disease, and eserine sulphate (one-quarter grain to the ounce) was instilled 3 times a day. The patient was kept under observation during the next two months, and although the development of the disease was gradual, it was constant. The eyeballs increased in size, the right eye being decidedly larger than the left. The corneæ became hazy, the pupils were slightly dilated, and responded sluggishly to light. Strange to say, the anterior

chamber was somewhat shallow. There was profuse and annoying lachrimation.

Johnson performed double iridectomy, and, although the iris prolapsed upon removal of the knife in both instances and the child was extremely restless, the result was most gratifying. About 6 months after the operation the parents brought the child for examination. She had no apparent trouble. The profuse lachrimation had ceased, and the eyes had not increased in size. She had apparently satisfactory vision. Upon examination the eyeballs appeared to have ceased the process of abnormal enlargement, the corneæ were clear, the tension was about normal, and the pupils were responsive to light. In the right eye there was a perfectly satisfactory coloboma; in the left eye there was a darkened scar at the limbus. At the point of incision the iris was incarcerated and the pupil distorted. Both pupils were responsive to light. The ophthalmoscopic examination was not entirely satisfactory; occasional glimpses of the fundus seemed to indicate the absence of disease; the vessels, fundus, and nerve-head appeared normal. The progression of the ocular growth was abated, and all of the indications forecasted future useful vision.

Miotics.—Both pilocarpin and eserine, being harmless and tending to reduce tension, are recommended, but are of little use when employed alone. As adjuncts to operations they are invaluable.

Constitutional treatment is indicated according to the child's general health, but is worthless as a direct curative measure.

Correction of the Myopia and Astigmatism.—The enlargement of the antero-posterior diameter of the globe in common (and, indeed, in excess) of the other dimensions produces a myopia, sometimes of such high degree as, in itself, to render vision worthless. Besides, the diseased cornea is likely to be astigmatic. Judging from the absence of mention in literature, these facts have been generally overlooked in the treatment of patients above 3 or 4 years of age. In the case clinically studied by myself, a child of 6 was considerably benefited by simple correcting concave lenses. In cases in which the media are still partially

clear and the nerve is not atrophic, if possible, the refraction of the eyes should be determined. This can be usually accomplished by retinoscopy and ophthalmoscopy, and substantiated by the test-lenses. Such refraction should be a useful later adjunct to operations.

Education at Blind Asylums.—If the child is semi-blind and well-developed mentally for its years, it should be placed in an asylum early, in order that its pliable mind and remaining active senses may be developed and its future life lightened before it becomes too morbid to take interest. With a good institutional training its usefulness in the world will not be entirely impaired.

Enucleation is demanded when the eye is painful, attains enormous proportions, becomes perforated or infected. *Evisceration, Mules' operation*, or any procedure intended to leave a movable stump in the orbit, would seem to be preferable to enucleation in cases other than those of panophthalmitis. By partially filling out the orbit they permit of a more symmetrical growth of the face.

CLINICAL STUDY OF A CASE OF BILATERAL HYDROPTHALMOS.

Personal History.—The patient was a boy of 6, born August 8, 1892, a healthy 8-pound baby. At about the third month the mother noticed what she called "a white scum" over both eyes. She denied all history of sore eyes in the boy. He had measles, varicella, and some minor ailments; but with no serious sequels. Since the age of 3 months, his eyes have appeared large and protruding and have attracted the attention of every trained observer under whose notice the boy has come. At the present time the boy is apparently in the best of health and is well-developed mentally.

Physical Measurements.—His height is 118 cm. (47 inches). His weight is 48 pounds; his chest-circumference is 56 cm. (22½ inches). The measurements of his head are extraordinary and significant of congenital abnormality in the skull. The head is oblong rather than oval. Its circumference is 55 cm. (22 inches)—little less than one-half his height. The distance from one external canthus to the other is 10 cm.; the distance between the internal canthi is 4 cm., making the palpebral fissure about 3 cm. on each side. The forehead is extremely high and broad; the distance from the nasion over the vault to the inion is 36 cm. (15 inches).

The lower part of the face is well-formed and normal in size.

There are no striking abnormalities of the nose, mouth, or throat. His teeth are carious and poorly formed, but not of the notched variety. The 6-year molars appear better formed than the temporary teeth. The palate is not highly arched, in fact it is well formed.

There are no deformities elsewhere on the boy's body, although



A Case of Bilateral Hydrophthalmos.

his forehead and scalp show scars, the result of many falls due to his defective vision, while playing with his companions. He eats well and sleeps soundly, and seems of good, but not especially cheerful disposition.

The lids are drawn apart in the accompanying photograph to better show the enlarged corneæ.

Family History.—The family history is good. There are 2 other children, a girl of 17 and a boy of 3, both of whom have normal eyes. The father is a robust, temperate man of 43, with $\frac{6}{8}$ vision in both eyes and strong accommodation. He says his wife has worn glasses for a number of years, but with them has perfect vision. He denies all history of syphilis and says there is no history of tuberculosis or alcoholism and little or no rheumatism in his or his wife's immediate family. There have been no club-feet, hare-lip, cleft-palate, or other marked congenital anomalies in the family.

Ocular Condition.—The sclera has the characteristic bluish-white appearance.

The corneæ measure about 16 mm. horizontally across the base (normal, about 12 mm.). The vertical diameter of the corneal base is about 15 mm. The corneæ are both maculated over their whole surfaces, but the opacities are particularly noticeable in the shape of a V of about 7 mm. base, extending from above downward to a point within 2 mm. of the center of the cornea. This would rather indicate that exposure was not the exciting cause of the opacity, as in such case the lesion would be below the center or confined to the palpebral fissure. The boy can close his eyes thoroughly and tightly, and his father claims that his eyes are always closed in sleep. Other than the corneal opacities, there are no signs of inflammation in the anterior eye. As in adult glaucoma, the cornea is quite lacking in sensation. Extending from the sclero-corneal junction around the upper half of the cornea is an irregular band of bluish-white opacity from 3 to 4 mm. in width, similar to arcus senilis except that it is continuous all the way to the scleral junction without any intervening rim of clear cornea, and it does not extend the entire circumference of the cornea. It is evidently denutritional in origin.

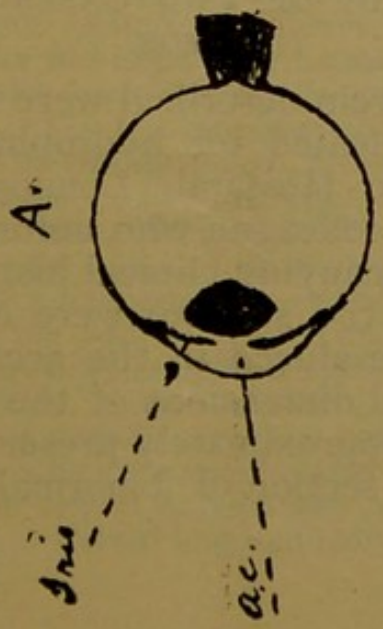
Ophthalmometric Measurements.—The radius of curvature of the right cornea is 8.9 mm. (7.8 mm. in the normal adult eye) and there is no astigmatism. The radius of curvature of the left cornea is 9.1 mm. and there is 0.50 D. of astigmatism at axis 60. Tension is fully + 1 in both eyes.

The iris is light-brown and very atrophic. The pupil is widely dilated (8 mm. in diameter) and responds only slightly to light-stimulus. The iris is not tremulous, showing that fixation of the lens was still firm. The anterior chamber appears very deep. As the surface of the iris is in a fairly even vertical plane, the approximate depth of the anterior chamber may be theoretically calculated from the known base-diameter and the radius of curvature—about 4 mm. in the right, and slightly deeper in the left eye.

Ophthalmoscopic examination was rendered most unsatisfactory by the maculated cornea. A bright red reflex was obtained by the indirect method, and now and then a peripheral retinal vessel could be made out, but no details of the macula or disc could be seen. There was a dark circular opacity floating in the vitreous of the left eye, which, at first was thought to be a permanent hyaloid artery.

COMPARATIVE DIAGRAM (ACTUAL SIZE) OF A NORMAL CHILD'S EYEBALL AND HYDROPHTHALMOS FROM INFLAMMATION AND FROM CONGENITAL MALFORMATION.

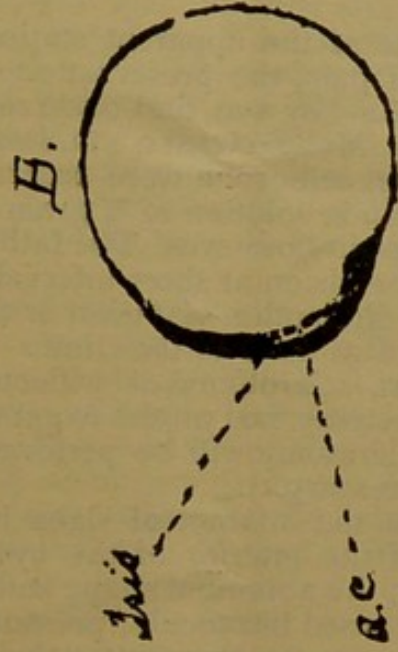
NORMAL INFANTILE EYEBALL.



Measurements in mm.

Base of cornea	12.1
Anteroposterior diameter	22.1
Vertical diameter	22.
Depth of aqueous chamber	3.1
Anteroposterior diameter lens	4.
Vertical diameter lens	7.2
Diameter of optic nerve	4.5

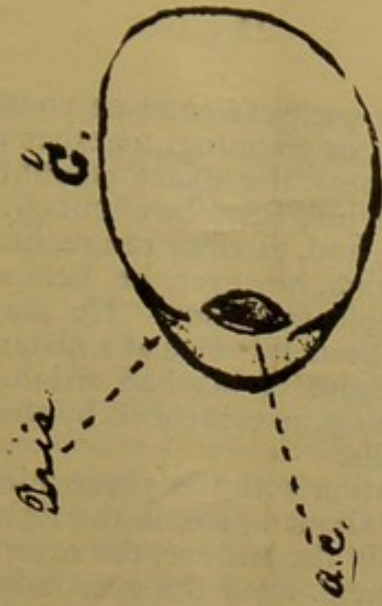
HYDROPHTHALMOS FROM INTRAUTERINE IRIDO-KERATITIS.



Case I.

16.2
28.3
25.1
.
.
.
.

HYDROPHTHALMOS FROM CONGENITAL ABSENCE OF CANAL OF SCHLEMM.



Case II.

14.2
30.1
22.
4.1
3.
6.2
.

Vision.—Owing to the patient's extreme youth and ignorance of the alphabet, figures, or counting, together with his childish indecision and nervousness, the exact amount of vision could not be obtained. It was judged to be about $\frac{1}{40}$. The boy could see a watch at 3 meters, and, at close range, he could tell colors and see fine objects which, however, he held very close to the eye, indicating his myopic condition. He plays with his little companions, recognizes them by voice at a distance, and by sight at about 4 meters. In spite of his bad vision, he has the boyish proclivities for romping, as evidenced by the numerous scars on his forehead from falls.

Retinoscopic examination with the plane-mirror showed a reversal of the shadow at about 14 cm. in the right eye, indicating a myopia of about 7 D.; in the left eye, the reversal took place at a distance of about 12.5 cm. from the eye, indicating a myopia of 8 D. According to these measurements the eyeballs were about 26 mm. long.

Treatment.—In view of the apparent stationary form of the disease, the absence of pain, the preservation of vision and the late stage at which the boy was first observed, operation was not deemed advisable. Myopic lenses, 1 D. less in strength than the retinoscopic findings indicated, were ordered and markedly improved vision. Eserin in solution of $\frac{1}{4}$ grain to the ounce was prescribed for daily use in both eyes. The father was instructed to methodically test the vision at short intervals and, at the first indication of decided diminution of vision or the appearance of pain, to bring the child at once to the clinic. In such event, if circumstances warrant, sclerotomy or iridectomy will be performed; perhaps iridectomy first on the worst eye, and if it fails to stay the disease, sclerotomy will be performed on both eyes and be repeated if necessary.

Conclusions.—From the absence of signs indicative of previous inflammation of the interior of the eye, the atrophic or maldeveloped iris, and the abnormal shape and size of the skull, I believe that the increased intraocular pressure in this case is due to congenital defective development rather than to introcular inflammation in uterus or in early infancy.

PATHOLOGIC STUDY OF TWO CASES OF HYDROPH- THALMOS.

The sections herein described were cut from infantile eyeballs, enucleated for hydrophthalmos at the Royal Ophthalmic Hospital, London. They were loaned to me by a colleague, who, unfortunately, could not furnish accompanying clinical histories.

The outlines of the sections were carefully traced by myself and reproduced in the accompanying diagram. The actual dimensions of the hardened eyeballs have been approximately preserved. The outlines of a vertical section of a normal child's eyeball

have been traced and reproduced for purposes of comparison. An attempt has been made to show the thickness of the cornea and iris in Case I. All the sections are median vertical.

Dimensions of the Normal Child's Eyeball.—Horizontal base of the cornea, 12.1 mm.; antero-posterior diameter of the eyeball, 22.1 mm.; depth of the aqueous chamber, 3.1 mm.; antero-posterior diameter of the lens, 4 mm.; vertical diameter of the lens, 7.2 mm.; diameter of the optic nerve just posterior to the lamina cribrosa, 4.5 mm.

Case I. (Fig. B. in the Diagram).—In this eyeball there were marked evidences of inflammation in the whole anterior segment. The iris was bound fast to the cornea in the greater part of its surface. The anterior chamber was almost entirely obliterated. Both the cornea and iris were thickened, and the cornea was markedly distorted. The sclera was considerably stretched and thinned. The lens was missing and, most likely, had been dislocated posteriorly and absorbed in the vitreous. The retina and choroid were detached and mostly disorganized. The scleral ectasia was not uniform, and the normal contour of the eyeball had not been maintained during the process of distension. In fact, the whole eyeball was disorganized and distorted.

Dimensions.—The antero-posterior diameter of the eyeball was about 28.3 mm. (22.1 mm. normal). The vertical diameter was 25.1 mm. (22 mm., normal). The horizontal diameter of the base of the cornea was 16.2 mm. (12.1 mm., normal).

From the macroscopic and microscopic findings, this case was evidently one of secondary glaucoma due to a congenital irido-keratitis, the elastic sclera of the infantile ball allowing total ectasia (hydrophthalmos), instead of limiting the extension to the region of the lamina cribrosa—the characteristic pathologic evidence of secondary glaucoma in adults.

Case II. (Fig. C in the Diagram).—The marked feature in this case was total absence of any indication of the canal of Schlemm. The eyeball was enormously enlarged in its antero-posterior diameter (30.1 mm.). The vertical diameter was about normal. The aqueous chamber was very deep—4.1 mm. (3.1 mm., normal). There was apparently no marked diminution of the iridic angle, which circumstance is confirmatory of Grahamer's observation. The lens was partially dislocated posteriorly and downward and was shrunken, measuring 3 mm. in its antero-posterior diameter (4 mm., normal), and 6.2 mm. in its vertical diameter (7.2 mm., normal). The iris was atrophic, being much thinner and smaller than is shown in the diagram. Judging from the space between the periphery of the lens and the ciliary processes, the zonule of Zinn must have been much stretched and consequently atrophied, allowing the lens to recede and drop downward. The cornea was increased in curvature and measured horizontally across its base 14.2 mm.; it was markedly sclerosed. The vitreous chamber was in a state of disorganization. The scleral ectasia was not symmetric, and the eyeball was much distorted longitudinally.

From the macroscopic and microscopic findings, there is strong reason to believe that the cause of this case of hydrophthalmos was defective development in the region of the filtration canals, rather than inflammatory obstruction or synechiæ.

SUMMARY.

1. There is an extraordinary state of confusion relative to the nomenclature, classification, etiology and treatment of these cases.

2. The proper name to employ to designate the condition is hydrophthalmos, as this indicates a general condition and not a special cause or local effect. By adhering to such a general name, no error in lexicography is committed, and future bibliographic research is materially assisted.

3. The disease is present at birth or occurs in early infancy, the inception being generally intrauterine.

4. The diagnostic symptoms are uniformly enlarged and protruding eyeball, insufficient lid-action, increased tension, sluggish and dilated pupil, atrophic iris which appears tremulous if the lens is luxated, peculiar bluish sclera, corneal opacity and anesthesia, deep anterior chamber, restlessness, ill-temper, constant rubbing of the eyes and other symptoms of local pain.

5. The cases generally classed under the name of hydrophthalmos or one of its synonyms are of different origin, and there is evidence in support of all of the following causes: (*a*) an intrauterine irido-keratitis, causing closure of the periphery of the anterior chamber; (*b*) congenital lack of development, either in separation of the iris from the cornea, or in deficiency of the filtration-angles in the neighborhood of the iridic angle; (*c*) a fetal serous cyclitis or uveitis causing excessive secretion with an accompanying diminution in the caliber of the veins of Leber's plexus and obstruction of the spaces of Fontana; (*d*) vascular disturbances producing arterial hypertension and trophic disorders.

6. Notwithstanding the several plausible theories, careful pathologic study seems to indicate that these cases may be divided into two classes:

- I. True hydrophthalmos, depending upon congenital defective development of the cornea, iris or filtration channels.
- II. Hydrophthalmos secondary to fetal intraocular inflammation, usually in the form of irido-keratitis or irido-cyclitis and keratitis, causing closure or obstruction of the iridic angle and the filtration-channels.

7. The prognosis of early operation in hydrophthalmos is far more favorable than the textbooks lead us to believe.

8. If a case of hydrophthalmos is recognized in the first few months of infancy and shows no diminution of progress under several weeks of palliative treatment with iodids, mercurials and miotics, repeated paracentesis should be tried. These failing, a broad iridectomy should be performed at once on the worse eye, unless the eyeball is very large, when repeated sclerotomies should be substituted. Sclerotomies failing in a medium-sized eyeball, iridectomy ought to be tried.

9. The earlier the operation the better will be the result. Late operations are dangerous because of the loose or dislocated lens, tenuity of the membranes, and disorganization of the vitreous. Prolapse of the vitreous and intraocular hemorrhage are the special dangers. The more patulous the filtration canals, the more likelihood there is for successful operation.

10. Miotics should be invariably used as adjuncts to operation.

11. The increase of the antero-posterior diameter in hydrophthalmos produces myopia, and the associate corneal disease is likely to cause irregular curvature-astigmatism. In cases giving evidence of useful near vision, whether operated upon or not, an attempt at correction of the refraction by retinoscopy and ophthalmometry, with confirmation by the test-lenses, is strongly advised.

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