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MEGALOCORNEA

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Submitted in Partial Fulfillment for the Degree of
Doctor of Medicine

College of Medicine, University of Nebraska

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I. INTRODUCTION

Although megalocornea is a well established clinical entity, there are still a number of controversies and areas of confusion. Etiology is very much an open question. Genetic and other studies suggest that megalocornea may be more than one disease form. The relation of megalocornea to certain other abnormalities of the anterior segment and indeed to generalized syndromes remains some what clouded. For example, the debate as to the relation of megalocornea and abortive hydrophthalmia once thought entirely settled has been reopened within the last decade.

The purpose of this paper is to discuss with some completeness the pathologic entity that is now commonly known as megalocornea. The subject matter is sufficiently limited that it is possible to review rather completely the literature on the subject. There is a fairly large foreign literature which I have gone over somewhat superficially relying heavily on the English sources.

My own interest in this subject is something of a personal one since my own corneas are pathologically

enlarged in association with a juvenile or late congenital form of glaucoma.

II. HISTORY

In 1869 von Muralt in his Inaugural Dissertation (Zurich) first suggested that "Cornea globosa semper pellucioda" (megalocornea) was not the same as congenital hydrophthalmia (buphthalmos). Horner and Michel, in 1889, differentiated "keratoglobus pelluciodus" from "keratoglobus turbidus". These authors also noted keratoglobus pellucidus was associated with a familial tendency, refractive error and disposition to "cataract and that slightly tremulous." They noted no increase in intraocular tension. These observations are remarkably close to the present day position.

Terson in France, performed the first successful cataract extraction on a case of megalocornea (1888). There continued to appear reports largely on the German literature of cases of large corneas unassociated with increased tension or other findings of glaucoma. Much confusion remained however and the general consensus (Axenfeld, 1905 and Collins 1913) was that these cases represented arrested or abortive congenital hydrophthalmia. With the publication of Kayser's pedigree of 17 cases in 1914 and Groenholm's pedigree of 13 cases in 1921, both showing a sex-linked recessive mode of in-

heritance, megalocornea was established as a separate clinical entity. Collins, in 1920, noted in print that his case of 1913, must rather have been megalocornea than an atypical hydrophthalmia. Seefelder (1916), Staehli (1914), and Kestenbaum (1919) all published accounts supporting megalocornea as a separate condition. Despite these publications some perplexity remained as late as 1930, when Doggart used megalocornea and buphthalmos synonymously.

A milestone was reached with Vail's detailed report (1931) in which he reviewed the world literature summarizing some 69 well studied cases. He demonstrated convincingly the difficulties encountered with cataract surgery in the face of megalocornea. From the time of Vail too, we note a general shift of authority in this field as in so many others in that era from the German to the English speaking world. Duke-Elder (1938) in Vol. II of his "Textbook" and Anderson (1939) in his monograph on Hydrophthalmia included important reviews on the subject.

Since then, scattered reports have continued to appear from time to time. Smillie (1955) brought the megalocornea-cataract literature up to date with 10 more cases including two of his own. Reports by

Hamilton (1951), Trautman (1952), Oppel (1957) and Malbran and Doods (1960) have reopened the question or connection between megalocornea and hydrophthalmia, though being careful to preserve in the main the now classical notions of Vail, Duke-Elder and Anderson.

III. DEFINITION AND DESCRIPTION

Megalocornea is generally described as a hereditary sex-linked recessive bilateral developmental anomaly found almost exclusively in males. The most striking clinical observation and the one most easily determined is of course the diameter of the cornea. Oppel (1956) states the average corneal diameter is 11.66mm. ±1.32 mm. A cornea must exceed 12.98 mm. or for practical purposes 13 mm. to be considered enlarged. He denoted megalocornea as an enlarged cornea associated with degenerative changes in the anterior segment of the eyes. He distinguished macrocornea which is a normal physiological variant. This is somewhat at odds with Anderson (1939) who stated the megalocornea is "a completely healthy eye in a healthy person". Friede (1933) agrees that 11.66 mm. is the average corneal diameter but makes his distinction thusly:

1. Normal corneae-11.0 to 12.0 mm. (horizontal diameter)
2. Microcorneae-5.0 to 11.0 mm. (horizontal diameter)
3. Macrocorneae (physiologically large) - 12.1 to 13.0 or 13.5 mm. (horizontal diameter)
4. Megalocorneae-11.0 to 18.0 mm. (horizontal diameter)

In any case there is agreement that the diagnosis of megalocorneae must depend on observations other than corneal diameter alone.

Over the years a confusion of terms has gradually arisen in the literature. The designation macrocornea has already been disposed of. Other terms put forth by various authors such as Keratoglobus, Keratomegalia, gigantophthalmus and anterior megalophthalmus are in effect synonymous with megalocornea. The first two are self-explanatory. The latter are based on their authors' ideas as to the true nature of the disease condition. Seefelder (1914) proposed gigantophthalmus in the belief that the enlarged cornea was actuallyaa proportional enlargement of the entire eyeball. It was generally felt by most authors that this was not the case. But the question was not settled until Kayser (1932, 1936) showed histologically the posterior segment to be normal in all respects.

Vail's term of anterior megalophthalmus enjoyed somewhat more vogue, the contention being that all the anterior structures were large (lens, zonule, iris, ciliary body). Rosen (1945) very succintly pointed out that this is not a true enlargement but rather a "stretching" in order to "stay with" the increased

limbal circumference. This stretching affects principally the iris (atrophy) and the zonule (weakening) giving rise to many of the accepted diagnostic criteria. The cornea is enlarged but not particularly stretched or rendered thinner although Dohlman and Larsson (1958) and Kayser (1936) report cases showing central thinning of the cornea.

The clinical characteristics of megalocornea (taken mainly from Anderson) may be summarized under the following headings:

1. Cornea. First there are the enlarged but healthy corneae varying from 12 to 18 mm. and most generally exceeding 13 mm. Corneal opacities, unless due to injury or secondary ulceration, are absent. There are no tears in Descemet's membrane. Melanosis corneae or Krukenberg's spindle may be present. Embryotoxon (areus juvenilis) or gerontoxon (arcus senilis) is frequently noted. The corneal radius of curvature is normal or less than normal, ranging according to Anderson from 7.0 mm. to 8.5 mm. and averaging 7.5 mm. which is also the normal average.

2. Refraction. Refractive errors are frequent with all types represented but astigmatism generally with the rule is unusually common. In Vail's series

out of 92 eyes, 24 had astigmatism with the rule and against. Vision is otherwise unimpaired unless secondary events occur.

3. Corneo-scleral junction. The limbus shows no widening as in hydrophthalmia. The corneo-scleral groove is sharply defined.

4. Sclera. This is characteristically normal even near the limbus where stretching occurs in hydrophthalmia.

5. Anterior chamber. This structure is unusually deep (4-8 mm). The absence of this striking feature in large physiological globe aids in the differentiation from megalocornea. The chamber is deep in advanced cases of hydrophthalmia.

6. Iris. The iris shows stretching and atrophy of its stroma. As a result of the weakening of the dilator muscle miosis is frequently present. Dilatation is often sluggish. As Rosen (1945) points out, the miosis may be relative owing to the large area of the iris. The sphincter is less affected because the atrophy seems to occur most markedly at the periphery. Iridodonesis is commonly present.

Krukenberg's spindle is not necessarily congenital and is thought by Anderson to represent scattering of

iris pigment.

Gonioscopy has not been reported by most authors. Detailed reports so far as I can find are those of Troncoso and Givner (1936), Rosen (1945), and Malbran and Dodd (1960). These and a few others are agreed that the iridocorneal angle is widened, often markedly. Other details are found in a later section.

7. Lens. As a result of stretching of the zonule the lens may be tremulous, subluxated or completely dislocated. The lens itself is of normal size although Klar (1940) reports extracting a cataractous lens from a man with magalocornea that weighed 340 mg.

The lens appears to have a strong tendency to opacities. Vail reported 27 cataracts out of 69 well documented cases. Tables I and II show 12 out of 28 more recent cases. It is interesting too, to note the large number of cases age 50 and below (9 out of 12). Vos suggests that this increased incidence of cataract is due to the disproportionate uvea.

Remains of pupillary membrane are not uncommonly found on the lens capsule.

8. Other Ocular Tissues. These are normal. There is no cupping of the optic disc. There is complete absence of signs of glaucoma including increased ocular

tension. In fact hypotension is fairly frequently noted. There were 5 patients with tensions below 10 mm of Hg. in Vail's series. Kayser notes 3 cases and Troncoso, Rosen and Peters (1925) each note one.

Anderson states that,

> "when hypertension is found with megalocornea it occurs late in life. It may be associated with either obstruction at Schlemm's canal by pigment, or sclerosis of the meshwork, or with subluxation of the lens."

Vail describes the disease by these widely known and accepted criteria:

Major signs--

1. Enlarged transparent corneas.
2. Hereditary and familial incidence.
3. Bilateral and almost exclusively male.
4. Absence of all evidence of glaucoma.

Minor signs--

1. Deep anterior chamber.
2. Iridodonesis.
3. Atrophy of iris stroma and miosis.
4. Embryotoxon or gerontoxon.
5. Dislocated or tremulous lens, frequently cataractous.
6. Melanosis corneae or Krukenberg's spindle.
7. Visible nerve fibers on cornea.

8. Remains of pupillary membrane.

9. Sharp corneoscleral margin.

Vail analyzes his minor criteria as follows:

Based on 69 cases--

1. Embryotoxon or gerontoxon 24
2. Krukenberg's spindle 4
3. Persistant pupillary membrane 8
4. Irododondesis and tremulous lens 42
5. Nerve fibers in cornea 5
6. Lens opacity 27
7. Deep anterior chamber 27
8. Irregular pupils 16

Based on 28 cases of tables I and II in this paper--

1. Embryontoxon or gerontoxon	.2.	8	10
2. Krukenberg's spindle	.0	1	
3. Persistant pupillary membrane . . .	0	3	
4. Irododonesis	5	8	13
5. Tremulous or subluxated lens . . .	.2	4	
6. Nerve fibers in cornea	.0	2	
7. Lens opacity (cataract)	4	8	12
8. Deep Anterior chamber	.10	18	28

The above lists, of course, have no statistical value but are useful in making certain comparisons. I should like to point out that this shows the type of cases published since Vail are very similar to those previously reported. Both lists indicate that the most prominent minor criteria are (a) embryotoxon, (b) irodonesis, (c) tremulous lens, (d) cataract and (e) deep anterior chamber. It is difficult to say whether these

are noted most often because they are more striking or truly more representative.

IV. DIFFERENTIAL DIAGNOSIS

The differential diagnosis of megalocornea is limited to two conditions lying in opposing directions. The distinction between megalocornea and microcornea (the benign anatomical variant) has already been discussed. In the other direction it is necessary to distinguish megalocornea and hydrophthalmia, an unquestionably pathologic state.

Hydrophthalmia is synonymous with the terms congenital glaucoma and buphthalmus. With Anderson, I prefer the word hydrophthalmia as being more descriptive of the actual condition.

Until the early part of this century as noted in the history, megalocornea was felt by most writers to be, in fact, a limited variant of hydrophthalmia. Seefelder (1916) listed these reasons for believing that megalocornea was not a result of increased tension;

1. The absence of corneal opacities and particularly tears in Descemet's membrane.

2. Though the anterior portion of the eye is enlarged, the limbus is not widened.

3. The corneo-sclerotic groove is normally defined.

4. The normal appearance of the sclera even in the region of the anterior chamber, where it would appear much more stretched.

5. The absence of any excavation of the optic disc.

6. The absence of functional disturbance.

7. Relatively high regular astigmatism. In hydrophthalmia it is usually irregular.

8. The corneal radius of curvature is normal or less than normal. In similarly enlarged hydrophthalmic eyes it is usually much greater, giving the cornea a flattened appearance.

9. Normal tension.

10. Bilateral symmetry. In bilateral hydrophthalmia usually a considerable difference is found.

Kestenbaum (1919) published a summary of the differential points between the two conditions. Here they are as modified by Duke-Elder (1938).

	Megalocornea		Hydrophthalmia
1.	Vast majority males	1.	5 males to 3 females
2.	Eyes bilaterally symmetrical almost invariably.	2.	35% unilateral
3.	Normal tension	3.	Tension increased.
4.	No corneal opacities including tears in Descemet's membrane.	4.	Opacities and tears common.

5. Normal or increased corneal convexity. Astigmatism commonly with the rule.	5. Decreased corneal convexity. Astigmatism against the rule.
6. No gross malformations of anterior chamber angle.	6. Gross abnormalities of angle.
7. No cupping of disc.	7. Frequent cupping of disc.
8. Familial--very common.	8. Rarely familial.
9. Little functional defect except refraction.	9. Much functional defect.

Were all diagnoses this definite medicine would be simple indeed! Unfortunately even the diagnosis of megalocornea is not this easily established, the vast majority of writers to the contrary. There is a condition variously known as arrested hydrophthalmia, hydrophthalmia sanatus and recently (Malbran 1960) as incomplete congenital glaucoma, which is very similar to megalocornea. Until 1914, as previously noted in this paper, no distinction was made. After this time the distinction was very emphatic though an occasional writer clung to the old view.

Hamilton (1951) in a rather sketchy article, mentions 15 cases, 7 or 44% of which were females. However, heredity was established in only 4 cases (all male and compatible with sex-linkage). Two of the females were affected unilaterally. Four cases (Hamilton does not say which 4) developed chronic glaucoma in later life. Hamilton's attempt to establish a relation be-

tween glaucoma and meglocornea falls through poor documentation. In 1955, Trautman notes a father with megalocornea (see Table II) and a son with bilateral buphthalmos. There is a family history of megalocornea but no pedigree given. Trautman concluded that megalocornea and buphthalmos are related. Oppel (1957) cites a similar case of a father with megalocornea (see Table II) and a daughter with bilateral buphthalmus. Oppel emphasized that there are no objective criteria to judge between megalocornea and arrested hydrophthalmia.

Malbran and Dodds (1960) may have succeeded in giving us just these criteria. After Kluysken's (1950, 1955) they accept this classification of congenital glaucoma:

1. Complete congenital glaucoma--early onset (at birth or infancy).
2. Complete congenital glaucoma--late onset (childhood or adolescence).
3. Incomplete congenital glaucoma--(tension normal).

Kluysken's criteria are as follows:

Pathognomonic of congenital glaucoma taken together--

1. Diameter of cornea in excess of 12 mm.
2. Unusual depth of anterior chamber.
3. Abnormal persistance of mesodermal tissue. at the iridocorneal angle.

Secondary symptoms--

1. Presence of ruptures or tears in Descemet's membrane.
2. Opacities in the corneal parenchyma.
3. Hypoplasia of iris stroma.

Kluysken's considers increased tension as but an added symptom of exeptional gravity.

It is clear that the afore mentioned incomplete congenital glaucoma is very similar to megalcocrnea. Malbran and Dodd suggest that objectively the only difference is in the iridocorneal angle. They maintain that the diagnosis of megalocornea is justified only after gonioscopy reveals a normal angle. The angle alternations most characteristic of congenital glaucoma, noted in gonioscopy, are (after Malbran): persistance of mesodermal tissue, abnormally wide aperture of the angle and almost constant invisibility of Schlemm's canal.

The gonioscopic findings of Troncoso and Givner, and Rosen generally agree well here and would be considered by Malbran as cases of incomplete congenital glaucoma. On the other hand, cases of megalocornea have not been studied gonioscopically and, minus these findings, the cases of Malbran and Dodd fit beautifully the description of megalocornea, with the exception that familial patterns are poorly established or absent.

We are left in an unfortunate position. The strongest evidence that megalocornea is a definite entity lies in the genetic pedigrees. However, none of these

cases has been adequately studied gonioscopically. Dohlman and Larsson (1958) missed an excellent chance in that regard with their sex-linked pedigrees of 8 cases. On the other hand those cases that have been well studied gonoscopically either lack a hereditary pattern or one has not been demonstrated.

V. INHERITANCE

The most compelling argument for considering megalocornea a separate pathologic state lies in the area of genetics. In this regard the pedigrees of Kayser (1914) and Groenholm (1921) have been widely hailed. These I have included along with those of Gredig (1926) and Dohlman and Larsson (1958) at the end. Three of these pedigrees are consistant with a sex.-linked recessive inheritance mediated through the female and transmitted to the male. Kayser's pedigree shows one case of unilateral megalocornea. Groenholm's show two afflicted females both however, resulting from a consanguineous marriage of an involved male and a presumably carrier female. Gredig's also shows two involved females, but the mode of inheritance appears to be that of autosomal dominance. It is important to note that no case of hydrophthalmia has been found in any of these females. Of the 73 cases noted by Vail, only 6 were females (2 were from Groenholm's series and 2 from Gredig.) This wide disparity in itself is indirect evidence favoring a generally sex-linked pattern.

Of the cases cited in Table II, only two failed to give a suggestive family history; of these one was an orphan. The cases of Trautman and Oppel had children with buphthalmos. Trautman's case otherwise had a family history suggestive of heredity megalocornea. The cases of Klar and Smillie seem to indicate autosomal dominance, while the cases of Posthumus and Vos show sufficiently incomplete histories as to be compatible with either a dominant or sex-linked recessive pattern. The remaining cases all suggest the sex-linked recessive mode.

Malbran's 10 cases show wide differences in this area. First of all there are as many females as males (5 and 5). Secondly only 3 are recorded as having a family pattern. Two are sisters related to a third girl with complete congenital glaucoma of late onset. The other was a male who reported a son as having the same condition.

VI. ETIOLOGY

Much has appeared in the literature concerning the origin of megalocornea but very little has been established. Most authors have chosen etiological theories in support of their own pet views as to the nature of the disease. As a result many of the notions about causation are now of historical interest only. Most prominent of these are first, the view prevalent before 1914, that megalocornea was a result of early but arrested high tension; and second, Seefelder's idea of gigantophthalmos. Both of these ideas have been previously dealt with.

Staehli suggested a hereditary hyperplasia of the cornea followed by pigmented corneal changes, iris atrophy and luxation of the lens.

Reis (1920) and Friede (1923) proposed that megalocornea was atavistic in origin. This idea was prompted by facts summarized on the following table taken from Mann (1957):

	Diameter of Cornea		Diameter of eye
1. Human foetus	1	:	1.7
2. Adult human	1	:	2.1
3. Ape	1	:	1.6
4. Pig	1	:	1.48
5. Sheep	1	:	1.47

6.	Cat	1	:	1.2
7.	Frog	1	:	1.2

This graphically illustrates that the proportional size of the adult human cornea is the smallest in the vertebrate division although the foetal cornea is relatively similar to the others in size as the table shows.

Mann states her position on etiology as follows:

> "The posterior parts of the eye are not enlarged though the ciliary ring undoubtedly is, since tremulous iris and spontaneous dislocation of the lens occur. It is probable that this enlargement of the ciliary ring is the primary aberrance, the cornea being necessarily larger since the circumference of the limbus is greater than normal. If this is so, the defect is one of relative growth rates of the various parts of the optic cup. Normally the cup is at first bell shaped and only gradually changes to a more spherical form. If the early embryonic relationship of diameter of anterior opening of the cup to equatorial diameter remained too long, the result might well be a permanent increase in relative diameter of the ciliary ring and hence an apparent greater size of the whole anterior part of the eye. This theory would place the fault on the ectoderm of the cup and would make it a condition of temporary slowing of the growth rate of one part compared with another. This does not conflict too badly with the usual atavistic theory since both in the lower animals, in their embryos, and in the human embryo, the condition has its possibility foreshadowed."

VII. IMPLICATIONS

Megalocornea as I have tried to show is not a serious abnormality in itself and by most definitions is non-progressive. Yet it is not a benign condition as the following discussion shows.

The best established associated finding is the predisposition to cataracts reflected both in frequency of occurance and in generally early age of onset. The status of cataract surgery in megalocornea has been summarized by Vail (1931) and Smillie (1955). Vail reported useful vision resulting in 2/3 or 18 cases viewed while Smillie states success in 9 out of 10. Vail mentioned vitreous loss in 7 while Smillie notes 2 out of his series. These figures, while not statistical, indicate that cataract surgery has improved since 1931, but is more hazardous than in uncomplicated cases.

The general problem of association of megalocornea and congenital glaucoma has been discussed. There are not enough reports to state a definite relation between glaucoma in later life and megalocornea although Hamilton believes that there is.

There is one report of megalocornea in a set of

twins afflicted with craniosynostosis (Calamandrei, 1950) but this is likely only a curiosity.

Various authors (Vail, Duke-Elder, Anderson) note that megalocornea is "frequently" associated with Marfan's syndrome. Rosen (1945) has this to say:

> "---------in reviewing Rados' (1942) most excellent, exhaustive and comprehensive work on Marfan's syndrome in which every case in the literature is tabulated (over 200 cases), I have been able to find megalocornea in only three cases. In these three cases is included assumption that megalocornea and arachnodactyly are frequently associated. Thaden's measurements were 13 mm and 12.5 mm, respectively. This same case had been reported one year earlier as a case of arachnodactyly and megalocornea by another author, Fleischer. In reviewing this same list of cases of Marfan's syndrome five cases of microcornea were uncovered. It seems therefore, that megalocornea is not a commonly associated finding in arachnodactyly------".

VIII. SUMMARY AND CONCLUSIONS

1. Megalocornea as a clinical entity is reviewed in historical perspective.

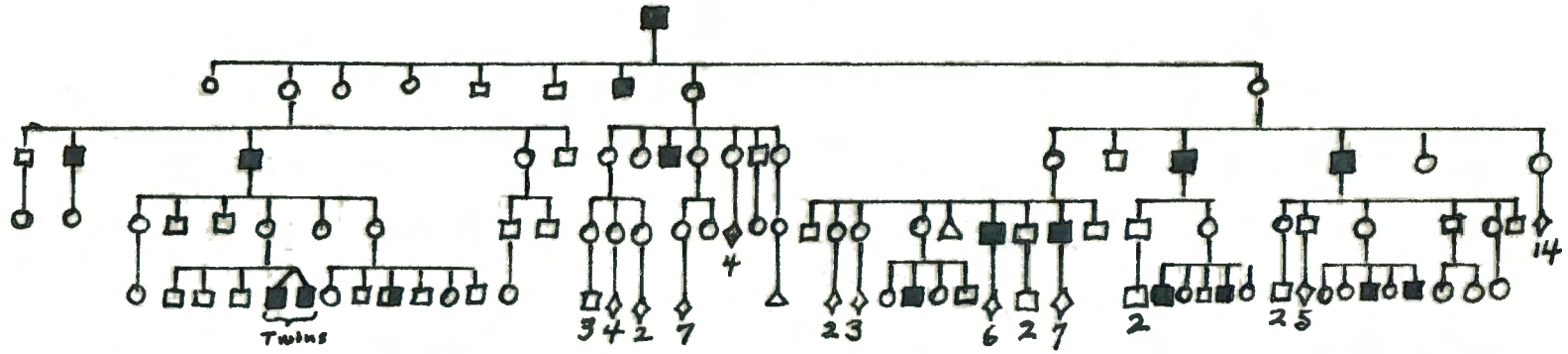
2.. The classically established form is a sex-linked recessive condition of enlarged cornea with frequently associated degenerative changes in the iris, lens and zonule.

3. The differential diagnosis of megalocornea once thought a closed question has been subjected to analysis in the light of recent articles. The differential is between megalocornea as defined above and incomplete congenital glaucoma (hydrophthalmus sanatus). It is pointed out that no final decision can be reached until cases of megalocornea with well established sex-linked pedigrees can be subjected to gonioscopic examinations as outlined by Malbran and Dodd.

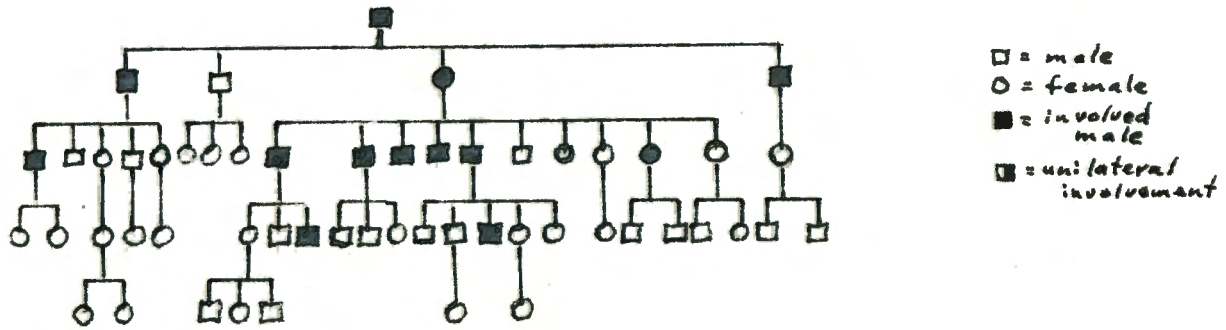
4. Etiology is discussed with final conclusions difficult to reach. The most prominent idea suggests that megalocornea is a result of atavism. This approach is emphasized in this paper.

Megalocornea is presented as a condition not entirely benign which is frequently complicated by correctable refractive errors and cataracts.

Genetic Pedigrees I.

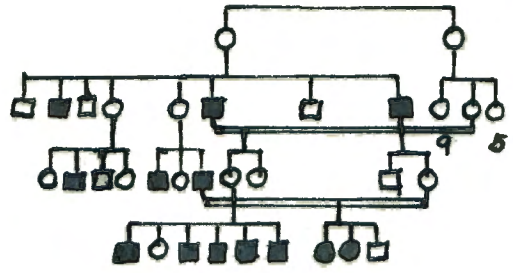


Kayser (1914). Pure sex-linked recessive pattern.

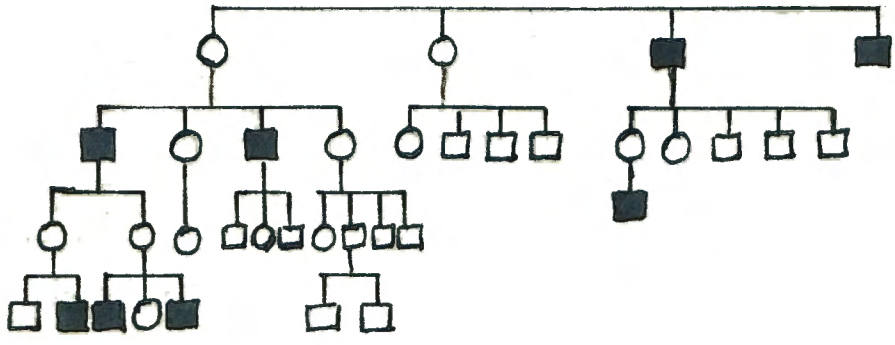


Gredig (1926). Pattern suggestive of autosomal dominance. Two females affected.

Genetic Pedigrees II.



Groenholm (1921). Sex-linked recessive pattern.
Double lines indicate consanguineous marriages.



Dahlman and Larsson (1958). Sex-linked recessive pattern.

Table I - Malbran's Cases

	Incomplete Congenital Glaucoma										Complete Late Congen. Glaucoma				
Malbran's pt. #	2	3	4	5	6	7	8	10	11	13	1	9	12	14	15
1. age	22	29	53	28	50	6	13	47	15	12	19	7	21	34	36
2. Sex	♀	♀	♀	♂	♂	♀	♀	♂	♂	♂	♀	♂	♂	♂	♀
3. Fam. History of Large cornea. B = Buphthalmus as well	+ B	+ B	–	–	+	–	–	–	–	–	+	–	?	+ B	+ B
4. Corneal Diameter O.D.	13	12	15	12	14	13	13	13	14	14	16	11	12	13	14
4. Corneal Diameter O.S.	13	12	15	12	14	13	12.5	13	14	gone	16	11	12	gone	13
5. Cornea Clear	+	+	+	+	+	+	+	+	+	+	+	No	+	+	?
6. Evidence of Glaucoma											+	+	+	+	+
7. Tension (Schiotz) O.D.	18	20	20	Hypo.	18	16	15	18	14	19	45	32	32	43	43
7. Tension (Schiotz) O.S.	17	18	21	16	18	17	16	10	16	–	50	37	49	–	27
8. Deep Anterior Chamber	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+
9. Iridodonesis			+	+	+		+	+							
10. Iris Atrophy	+	+	+	+	+	+					+		+	+	
11. Embryotoxon								+	+				+		
12. Tremulous or Dislocated Lens			+	+											
13. Cataract			+	+	+			+							
14. Krukenberg's Spindle															
15. Nerve Fibers on Cornea															
16. Pupillary Membrane											+				
17. Sharply Defined Corneasclerel Margin.															
18. Gonioscopy / Wide Angle	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+
18. Gonioscopy / Mesoderm	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+

Table II.

Review of more recent literature. Only well documented cases selected.	Davies	Trgmoso	Posthumus (Brothers)			Klaa (2 brothers 1 son)			Savin	Rosen	Natoff & Fonda	Vos (Half-bro. thru mother)		Trautman	Smith	Oppel	Dahlman & Larsson	Ryan
1. Age	66	28	27	28	39	43	13	39	58	26	51	51	30	3rd decade	50	29	60	39
2. Sex	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂
3. Fam. History S.L.=Sex-link D=Dominant B.=Buphthalmos fam.	S.L.	D. fam.	S.L. or D.	S.L. or D.	S.L. or D.	D.	D.	D.	S.L.	S.L.	S.L.	S.L. or D.	S.L. or D.	B.	D.	B.	S.L.	—
4. Corneal Diameter O.D.	13	13	14	13	14.5	15	13	15	14	16	16	14	13	14	17	13.25	16.5	15
O.S.	14	13.5	14	13.5	14.5	15	14	15.5	14	16.5	17	14	13	15	15	13.5	16.5	16
5. Clear Cornea	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	±	+
6. Tension (Schiotz) O.D.	16	8	Normal	Normal	25	Normal	Normal	Normal	Normal	9	25	Normal	Normal	15	16	Normal	Normal	20
O.S.	18	8			25					11	25			12	16			20
7. Deep Anterior Chamber	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+
8. Iridodonesis	+	+								+	+	+	+				+	+
9. Iris Atrophy		+	+	+	+				+	+		+	+				+	
10. Embryotoxon	+	+							+	+	+			+	+		+	
11. Tremulous or Dislocated Lens	+	+															+	+
12. Cataract	+				+	+			+			+			+		+	+
13. Krukenberg's Spindle										+								
14. Nerve Fibers on Cornea		+								+								
15. Pupillary Membrane		+	+	+														
16. Sharply Defined Corneoscleral Margin						+								+	+			
17. Gonioscopy – Wide Angle		+								+	+					+		
18. Gonioscopy – Mesoderm		+								+	?					?		
19. Evidence of Glaucoma																		

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