

LENS EXTRACTION IN MARFAN'S SYNDROME

BY

A. BENEDICT RIZZUTI, M. D.

Brooklyn, U. S. A.

In 1896 when Marfan (1) first described the syndrome that bears his name, no mention was made of the presence of any ocular manifestations. The syndrome is now considered to be a hereditary, bizarre clinical entity which can involve the skeletal, ocular, and cardiovascular systems in varying degrees.

Its true etiology is still speculative. Characteristically the affected individual is unusually tall, slender, with long arms and tapering elongated fingers and toes. Achard (2) in 1902 chose the term arachnodactyly to describe the spider-like aspects of the digits. Kyphosis, scoliosis and "pigeon breast" are frequent findings. The cardiac defects may be represented by valvular lesions, congenital heart disease and aortic aneurysms.

In 1924, Ormond and Williams (3) were the first to stress the importance of the ocular lesions, the most common of which are ectopia lentis, iridodonesis and miosis. These findings were present in approximately 50 per cent of reported cases Marfan's syndrome. Ectopia lentis is found in about 75 per cent of cases presenting this disease entity. The subluxated lenses may appear normal but usually show spherophakia, microphakia and varying degrees of lenticular opacities. The lenses are usually displaced superiorly in a symmetrical fashion. The syndrome as a rule is not usually detected until about the third or fourth year of life, when the demand for greater visual acuity increases. The syndrome occurs with equal frequency in both sexes.

In 1942, Rados (4) in a comprehensive report, compiled statistics of this syndrome in 204 cases published by a variety of authors. To date, reports of more than 300 cases of Marfan's syndrome have been recorded.

Many experienced surgeons are hesitant in attempting lens removal to improve visual acuity in this syndrome because of the high percentage of surgical difficulties and late complications that can occur. The elusiveness of grasping the abnormal pellet like lens, the great frequency of vitreous loss, and the ever

present danger of retinal detachment are now well recognized. Zeeman (5) pointed out that approximately 10 per cent of cases ended in spontaneous detachment of the retina. Surgical intervention was not a prerequisite in producing this complication.

In spite of these hazards, however, an occasional gratifying result may be obtained. For this reason the following case history is reported. Lens extraction was performed in the two eyes of the same individual who presented a typical clinical picture of Marfan's syndrome. The post-operative course was satisfactory and a visual acuity of better than 20/20 was achieved in each eye.

CASE REPORT

H. M., a white 30 year old male, was first seen on December 31st, 1954. The family history revealed that his father and one brother were also afflicted with Marfan's syndrome. Physical examination of the patient demonstrated the classical triad. Scoliosis and "pigeon breast" were very evident; the heart findings presented extensive valvular lesions. The ocular findings consisted of bilateral ectopia lentis, associated with a slight degree of iridodonesis.

After dilatation with 10 per cent neo-syneprine, both lenses appeared grossly clear and displaced inferiotemporally. The presence of free vitreous lying anterior to each lens was revealed by slit lamp examination. The corrected vision in the right eye was 20/80 with the use of -12.50 diopter sphere -1.75 diopter cylinder, axis 180. The corrected vision in the left eye was 20/100 with the use of -13.25 diopter sphere -2.00 diopter cylinder, axis 30. The patient was promoted to master electrician and demanded greater visual acuity hence operative interference was seriously weighed.

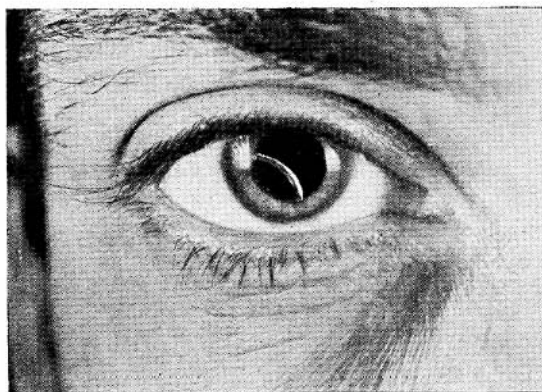
Lens extraction of the left eye was performed on January 3rd, 1955. The corneal section was made with a keratome and prior to performing an iridectomy



Fig. 1. Marfan's syndrome; note elongated, tapering fingers and chest deformity.

MARFAN'S SYNDROME

Fig. 2. Marfan's syndrome; clear lens subluxated inferior temporally, right eye.



superiorly a small amount of fluid vitreous escaped. The superior pole of the subluxated lens was grasped with a Verhoeff forceps and extracted intracapsularly with comparative ease and no further loss of vitreous. Air was introduced in the anterior chamber with guided pressure after firm closure of the corneo-scleral wound was obtained with the use of 5 corneo-scleral sutures.

The patient made an uneventful recovery and was given his final aphakic refraction on March 26th, 1955. Visual acuity of 20/16-3 was obtained with the use of +11.00 diopter sphere +1.00 diopter cylinder, axis 60.

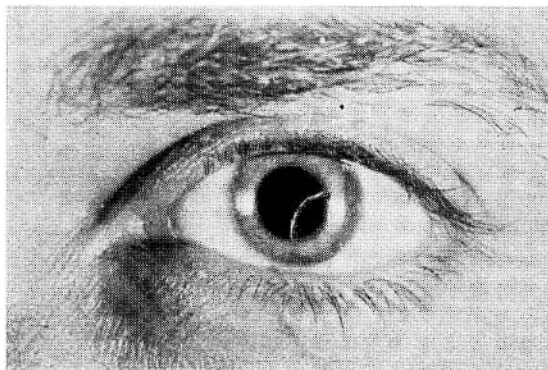


Fig. 3. Marfan's syndrome; clear lens subluxated inferior temporally, left eye.

Lens removal of the right eye was subsequently conducted on January 6th, 1956. After performing a wide iridectomy above, a loss of a small bead of semi-fluid vitreous resulted. The rather clear lens floated somewhat into the wound area and was easily extracted with a lens loop. The few adherent remaining zonular fibers did not offer any unusual resiliency. A slight degree of hyphemia was the only other surgical complication. The post-operative course, however,

was satisfactory. After the final refraction was performed on March 22nd, 1956 the visual acuity had improved to 20/16-1 with the use of a + 10.00 diopter sphere +1.75 diopter cylinder, axis 65°.

The patient was subsequently examined at regular intervals and at no time did he present any late surgical complications or diminution of vision. On October 5th, 1958 he expired while in the line of employment as a result of a heart attack.

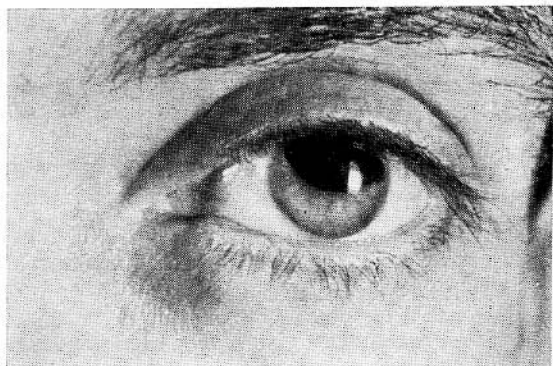


Fig. 4. Marfan's syndrome successful intracapsular lens extraction.

NON-SURGICAL CONSIDERATIONS

Surgical intervention as a rule should be postponed in arachnodactyly with ectopia lentis, if by means of refraction one can improve the visual acuity, either through the aphakic or phakic region of the pupillary zone to 20/70 or better. The refractive error through the phakic area at times may require a minus lens of 15 to 60 diopters. One should be aware of the possible presence of a congenital type of amblyopia with or without manifestations of strabismus. Myopia with astigmatism may be unusually pronounced and of lenticular origin. The subluxated lens across the pupillary zone can produce an annoying type of monocular diplopia and of necessity must be eliminated.

In cases where surgery is not feasible and when refraction does not improve the vision sufficiently, a weak mydriatic instilled daily may offer further improvement. Frequent tonometric and tonographic studies help rule out the existence of an underlying glaucoma.

SURGICAL INDICATIONS

1. Marked impairment of visual acuity.
2. Progressive lens changes.

3. Disturbing monocular diplopia.
4. Dislocation of the lens in the anterior chamber which may eventually lead to severe acute congestive glaucoma.
5. Dislocation of the lens in the vitreous resulting in frequent attacks of cyclitis and secondary glaucoma.

Biomicroscopic study should be carefully conducted of the iris, lens, vitreous and zonular area prior to contemplating any surgical procedure. This is best accomplished after dilatation with a mydriatic. The lens structure, extent and type of subluxation should be given careful consideration. The quality and position of any free lying vitreous, anteriorly situated to the displaced lens may lead one to alter the surgical procedure.

At times one is able to inspect the abnormal zonular area. It is a well recognized fact that although the zonule may be degenerated, the remaining adherent fibers can at times portray an unusual degree of resiliency elasticity. It is possible for a lens that eludes a spoon or where traction is lost with either the forceps or suction cup, to snap back with startling force and lose itself in the vitreous chamber. These factors and many others should be carefully weighed prior to considering any surgical intervention.

SURGICAL PROCEDURES

- I. Iris Surgery (optical improvement)
 - A. Sphincterotomy
 - B. Iridotomy
 - C. Iridectomy
- II. Lens Surgery
 - A. Discission (young individuals).
 - (a) use of one needle knife.
 - (b) use of two needle knives.
 - B. Intracapsular Extraction
 - (a) use of forceps.
 - (b) use of loop or spoon.
 - (c) use of suction.
 - (d) use of cautery.
 - C. Extracapsular Extraction.
 - D. Especial Cases.
 - (a) dislocation of lens in anterior chamber.
 - (b) dislocation of lens in vitreous.

IRIS SURGERY

No one particular surgical procedure should be universally adopted in surgery of a subluxated lens. Sphincterotomy or iridotomy are best performed in the lower nasal quadrant of the eye for optical improvement. A 3 or 4 mm. radial incision is usually found to be adequate in cases where the lens is displaced upward.

In cases of inferior subluxations a wide iridectomy superiorly at times produces a rather astonishing satisfactory optical result. These procedures should be attempted after proper visual evaluation following dilatation of the iris. It is often found, however, that these surgical procedures may prove to be disappointing for they are not easy to perform technically due to improper lens support and furthermore that they do not attack the main source of the difficulty.

LENS SURGERY

Young individuals respond better than adults to surgical intervention. Before the age of 25 years discission of the lens with the use of 1 or 2 needle knives can offer satisfactory visual results. The one knife technique is not recommended because of difficulty encountered in penetrating the usually small globular dislocated lens. This surgical procedure is difficult to perform in that the unsupported lens eludes the point of the knife.

Unless a wide rent is made in the lens cortex, the absorption of lens fibers are unduly delayed. Reese was of the belief that this phenomenon was an inherent characteristic peculiar to congenital subluxated lenses. Discission of the lens with the use of two needle knives as first described by Bowman in 1852 is still the operation of choice. It was popularized by Knapp⁶ and recently modified by Chandler⁷, Kravitz⁸, et al.

One needle knife is introduced through the sclera just posterior to the limbus at a point opposite the free border of the lens. The lens is pierced and held firmly in place. Another similar knife is introduced in the same manner a sector's distance away to puncture the lens capsule and cortex. The handles of the two knives are slowly approximated, the opening in the lens is enlarged and each knife is then cautiously removed in turn. Proper opacification of the lens fibers result but occasionally this procedure has to be repeated one or more times.

In the older age group intracapsular extraction with a wide iridectomy is the operation of choice. Free vitreous is frequently found to be present anterior to the lens and there may be some spill after making the corneal section. The proper iridectomy avoids the drawn up or hammock shaped pupil that usually follows.

It is possible to grasp the free border of the lens with a forceps to execute the proper extraction.

Gentleness and minimal amount of surgical manipulation helps prevent excessive loss of vitreous and usually leads to fewer post-operative complications. The lens spoon or loop when carefully used has its greatest value when the lens is seen to float rather freely towards the wound area. One must be careful of the zonular fibers which may show a live, elastic band response in cases where the lens slips from the spoon or the forceps. The lens may snap back with startling force into the vitreous chamber.

The hand or motor driven erisophake are often found to be inadequate in extracting the lens because of poor adaptation of the suction cup to the anterior lens surface in the presence of free vitreous. Barraquer⁹ suggested that in cases where the vitreous can be swept away with a Marten's hair brush that proper contact can then be made and the lens can thus be extracted by suction. The use of electrodiathermy for extracting the lens as advocated by Lacarrere¹⁰ has not been very popular. The Lacarrere electrodiaphake is used to pinion the lens. The instrument or electrome is released by means of a spring and enters the lens capsule and cortex to a depth of 3 mm. When the current is turned on, there results a coagulation of the lens protein which fixes the lens firmly and permits it to be extracted intact.

Dislocation of a subluxated lens in the anterior chamber may lead to acute secondary glaucoma and requires immediate extraction. In cases where recurrent dislocation of the lens in the anterior chamber occurs and then the lens slips back in position due to a relative intact zonule, Chandler⁷ performs a prophylactic peripheral iridotomy or iridectomy. By this means, pupillary block and secondary rise in intra-ocular pressure are avoided, which he believed was the cause for the lens to be dislocated in the anterior chamber.

When secondary glaucoma and recurrent attacks of cyclitis result, lens extraction is mandatory. In cases where the eye is quiet, it is wiser to avoid surgical intervention and keep the patient under careful observation. An aphakic correction at times can afford the patient useful vision for a considerable time. The use of a Hague Lamp which employs ultraviolet light is of much benefit in locating the position of the lens. The Verhoeff "floating technique" in which a stream of normal saline is directed into the vitreous chamber could be of much help in floating the lens upward so that it can be easily grasped and spooned.

CONCLUSIONS

1. Dilatation of the pupils in children under 3 years of age with defective vision should be a routine procedure to eliminate the possible presence of ectopia

lentic. After the proper preliminary study of the position of the lens and vitreous, the contemplated type of surgical interference should be carefully weighed.

2. The surgical results of lens extraction often prove disappointing because of associated ocular abnormalities.

3. In young individuals the operation of choice is dissection of the lens with the use of two needle knives, as originally advocated by Bowman. In older persons, intracapsular extraction with a wide iridectomy is the preferred operation. This may be accomplished with the use of a lens forceps, lens spoon or loop and rarely with the use of a suction cup.

4. Extracapsular extraction should not be attempted, for its results are exceedingly disappointing in Marfan's syndrome.

SUMMARY

The various surgical techniques in Marfan's syndrome have been reviewed and evaluated. A case report is illustrated in which two eyes were operated on in the same individual, one with the use of lens forceps and in the other using a lens loop. The results were unusually satisfactory. Vision of better than 20/20 was obtained in both eyes. The patient maintained useful vision and presented no post-operative complications after a follow-up period of 3 years. Eventually he expired of an acute cardiac distress while at work.

160 Henry Street

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