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Case Report Bilateral Crystalinian Ectopia Surgery In The Marfan's Disease In A 16-Year Child

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Summary: *The authors report a case of bilateral lens ectopia in Marfan's disease, in a 16 years old female child, who underwent surgery for the first time at the University Clinics of Lubumbashi, by phacophagy technique associated with a anterior vitrectomy by the pars plana. This observation helps to draw the attention of the community scientist on early surgery in case of bilateral lens ectopia in children to prevent amblyopia.*

Keywords: *lens ectopia, Marfan, phacophagy, vitrectomy*

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I. Introduction

Marfan syndrome is an inherited connective tissue disorder, transmitted according to the dominant autosomal mode with variable penetrance and expressiveness. The genetic anomaly is caracterised by a generalized dysplasia whose mesenchymal manifestations are observed in tissues and organs derived from the mesoderm as eye, skeleton and aorta mainly (1). Her prevalence ranges from 1/5000 to 1/20000 (2) with a rate of new mutations estimated at around 15% (3,4).

Ocular manifestations of Marfan syndrome, the most common of which is lens ectopia, are seen in 50 to 80% of the cases according to authors. These events often attract attention, leading the ophthalmologist to diagnose disease (4,5).

Patients with lens ectopia complain of a decrease or fluctuation of visual acuity and monocular diplopia (6). All directions are possible for lens ectopia, the most frequent direction is in super-temporal (7).

Enophthalmia, especially in children with severe disease, is related to the reduction even the absence of retrobulbar fat. The decline in visual acuity among young people is linked to an anomaly of refraction with development of an amblyopia which is most often bilateral. In the syndrome, patients have extreme refractive errors from moderate to strong myopia and astigmatism (6). We can also note a keratoconus (7).

Histologically, on a normal eye, the zonal fibers and the part of the capsule, where the zonule takes attachment, are rich in fibrillin. The percentage of fibrillin decreases gradually towards the center of the lens ectopia, which is free of fibrillin. In patients with Marfan syndrome, there is a low concentration of fibrillin at the equator level. Zonular fibers are rare, thin, sometimes broken and do not respect the parallelism of their alignment by randomly arranged(6).

Patient and observation

We present in this work a case of a 16-year-old patient who consulted University clinics of Lubumbashi for far vision loss about 3 years ago as well as the impossibility of putting the shoes because of deformation of her feet. In her past medical history, she is the eldest of a family of 7 children including 2 girls and 5 boys. No notions of tall person in his family or in his extended family. Objectively, oculomotricity was conserved. Far visual acuity without correction in both eyes was for hand movements at 50 cm not improvable

by corrective lenses. Slit Lamp Examination has shown an ectopia of the opacified bilateral super-temporal lens. The dilated fundus was hardly accessible to both eyes. The pediatric examination revealed an elongated morphotype (180 cm: picture1) with a weight of 53 kg, a rounded face, base of the nose wide and high, short columella, ears detached, slight microtia. The palate is hard of appearance and normal consistency (picture 2). The extremities which have tapered fingers (dolicodactylia) with ligament hyperlaxity (picture 3). The thumb and wrist sign are present (picture 4). The feet are flat (clinodactyly of 2nd and 3rd toes: picture 5). The chest x-ray revealed a slight dorsal scoliosis. Heart doppler ultrasound showed grade 1 aortic insufficiency (moderate dilatation of the initial aorta). The patient underwent surgery for lens ectopia opacified in both eyes consisting of anterior extraction of the lens (aspiration crystalline masses using a cannula) associated with implantation of a lens of 18 diopters (Artisan Implant) on the anterior surface of the iris to the right eye associated with it an iridectomy at 12 o'clock to prevent secondary glaucoma and implantation of a lens of 18 diopters in the posterior chamber of the left eye.

II. Discussion

Marfan syndrome is a rare disease that affects the connective tissue of the whole of the body, lens ectopy is the most common ophthalmological manifestation and is often bilateral; cataract occurs early in evolution (8). Such is the case of our observation which required surgery in both eyes seen that the ectopia of the opacified lens was bilateral.

Several surgical techniques have been described for ectopic lens in children: intra capsular, extra capsular, lens extraction with implantation of the lens in anterior or posterior chamber. In postoperative, we noted a complication frequently encountered in the surgery of lens ectopia in the Marfan syndrome which is the irregularity of the pupil. This was also observed by Necip et al. But these authors also report other complications that we have not found on our patient such as the decentralization of the implant, cystoid macular edema, uveitis (9).

Functional success was obtained more in the right eye where visual acuity was 5/10 with fault and where the implantation of the lens was made in the anterior chamber and fixation scleral following the irregularity of the pupil; in the left eye, visual acuity was fingers counting. Vadalà et al. (10) report that intraocular lens implantation using the scleral fixation technique is the first choice for patients with Marfan, because it reduces the complications of the decentralization of the lens.

III. Conclusion

Marfan syndrome is a rare pathology that is often accompanied by bilateral lens ectopia, several techniques have been proposed for children.

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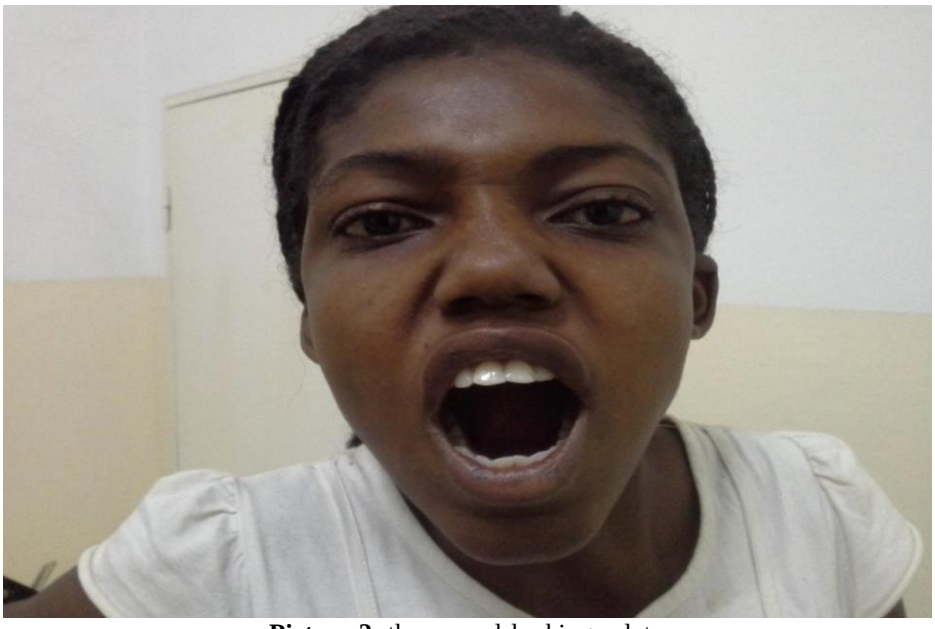
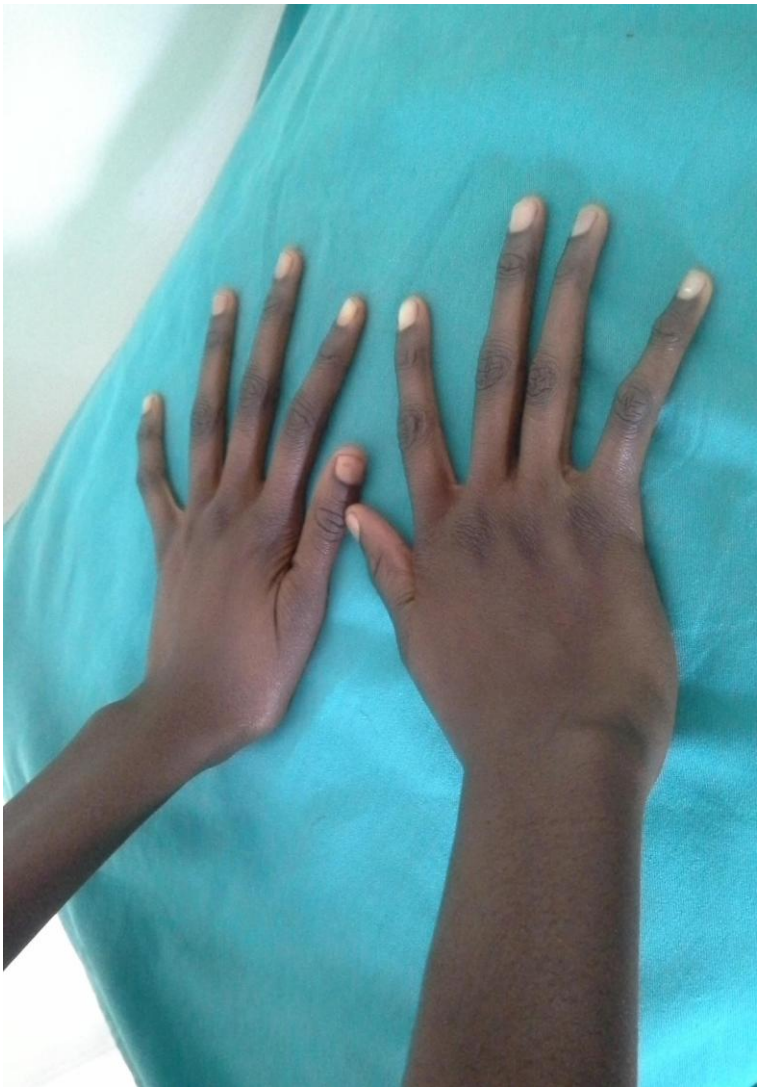
Picture 1: Size 1, 80 m



Picture 2: the normal-looking palate



Picture 3: Tapered fingers

Picture 4: thumb sign

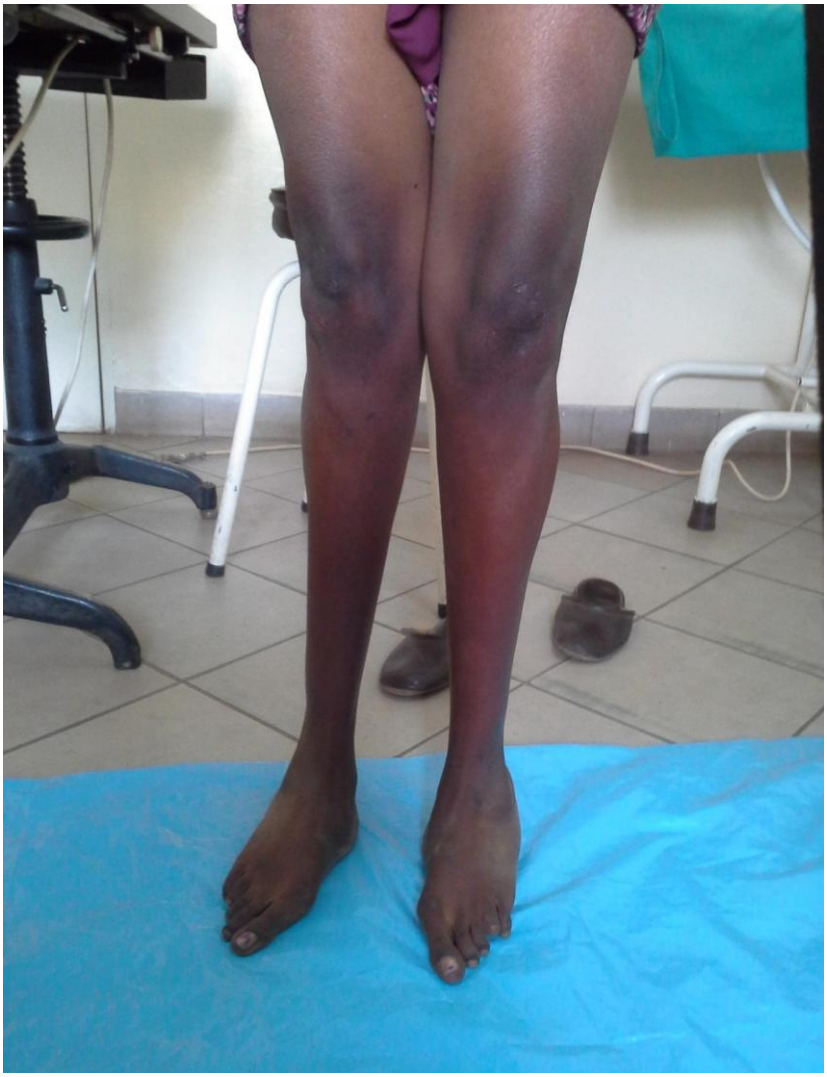


Photo 5: clinodactylie

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