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# Rare Presentation of Bilateral Filiforme Adnatum in a Syndromic Newborn

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## Introduction

Ankyloblepharon filiforme is a very rare congenital anomaly, characterized by partial adhesions of upper and lower eyelids. We report a case of case of bilateral AFA, associated with trisomy 18 (Edward syndrome) and a heart anomaly.The adhesions were successfully released by vannas scissors in OT without any anesthesia. It is extremely important to excise the adhesions early on, because of the risk of occlusion ambylopia.The purpose of this report is to make all general ophthalmologists aware of this abnormality and the fact that excision is very simple and can easily be done by a general ophthalmologist.

**Keywords:** Ankyloblepharon filiforme adnatum, Edward syndrome, occlusion amblyopia

## Case Report

A four days old infant was referred to ophthalmology department with complaints with the baby not being able to open the eyes since birth. The baby, weighing 2.5 kg was born at 38 weeks gestation with normal vaginal delivery. The mother denied any history of X ray exposure, drugs or any significant illness during pregnancy. The previous siblings, four year old and 6 year old were normal healthy. There was no history of consanguinity or congential defects as per history.The infant had been delayed for referral as already he was being investigated for multiple systemic conditions and was finally diagnosed as trisomy 18 (Edward syndrome) by chromosomal studies and also had multiple congenital cardiac anomalies. He was unable to suck, so had to be fed by a nasogastric tube. We diagnosed him as a case of ankyloblepharon filiforme adnatum(AFA)(photos to follow 2 in number)

On general inspection, clinical features of note were the micrognathia,low set ears, oddly shaped head and high arched palate. Relevant orbital findings included hypo plastic supraorbital ridge, mongoloid slant and mild epicanthus. On ocular examination, the baby was noted to have bilateral fused eyelids, with further detailed examination showing multiple bands of skin measuring 3mm in length and about 1.5 mm in breadth between upper and lower eyelids. The bands were divided by a vannas scissors without any anesthesia. Minimal bleeding was encountered. Post excision the external examination, motility and fundus were found to be unremarkable.

**Picture 1**- Bilateral adhesions of lids with bands

**Picture 2-** Bilateral bands in different view

**Picture 3-** Separated eyes lids after simple cut by vannas scissors

## Discussion

AFA is a rare benign congential anomaly, usually associated with multiple congential malformations, suggesting a genetic basis with variable penetrance. However, sporadic cases have been reported as well. There is no sex predilection described. Developmentally, the embryological defect is inability of eyelids to separate, which normally is complete by seventh month of gestation. The histology of connective tissue strands has been shown to consist of central vascularized core surrounded by stratified squamous epithelium.Rosenman in 1980 classified AFA into four groups-isolated, associated with multisystem syndromes, with ectodermal syndromes, with cleft lip/palate. Pathogenesis of this condition is debatable-intrauterine inflammation, trauma, but aberrant growth, as a result of interplay of temporary epithelial arrest and rapid mesenchymal proliferation, allowing union of lids at one or several points. In trisomy 18, it has been postulated that there is an abnormal cellular proliferation and hyperplasia of ectoderm and mesoderm on the lid margin. Known syndromes associated with AFA are Hay Well's syndrome, cleft lip/palate, popliteal pterygium syndrome and ectodermal dysplasias.Few cases have been described in associated with trisomy 18 also.

## Conclusion

AFA is a very rare anomaly but an ophthalmologist might encounter it, once in a lifetime. It is very important to diagnose it and suitably correct the deformity. The surgery is extremely simple and needs no special skill. Purpose of this report is to make a general ophthalmologist aware about the condition and do the surgery quickly as delaying might increase the chances of occlusion ambylopia.Also, seeing it, should the ophthalmologist to start investigating for systemic investigations too with the help of the pediatrician.