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## Long term follow up of mucous membrane grafting for cicatricial ectropion in Ichthyosis: A case report

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## ABSTRACT

**INTRODUCTION:** Ichthyosis is a group of keratinizing diseases characterized by scaly and dry skin. One of the ocular complications associated with ichthyosis is cicatricial ectropion which often results in exposure keratopathy and eventually corneal scarring.

**PRESENTATION OF CASE:** In this report we are presenting a 21-year-old female who is known to have ichthyosis-related bilateral lower lid cicatricial ectropion, which was managed with Mucous Membrane Grafting (MMG) six years ago.

**DISCUSSION:** Despite the serious complications of ichthyosis-related eyelid malposition such as cicatricial ectropion, until present no single way has been standardized to treat the ectropion in these patients. MMG has been previously described, but up to our knowledge this is the first reported long term follow up of MMG offered for ichthyosis-related cicatricial ectropion.

**CONCLUSION:** We strongly recommend MMG as a promising surgical treatment option offered for ichthyosis patients with cicatricial ectropion providing good outcome and acceptable cosmetic results.

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## 1. Introduction

Ichthyosis is a skin disease associated with many ocular conditions including cicatricial ectropion, in which patients suffer from ocular surface exposure consequences. Despite its serious complications, until present no single way has been standardized to treat ectropion in patients with ichthyosis.

## 2. The case

A 21-year-old girl who was known to have lamellar type ichthyosis, developed bilateral lower lid cicatricial ectropion related to her condition. She underwent Mucous Membrane Grafting (MMG) six years ago at King Abdulaziz University Hospital, Riyadh for management of anterior lamellar severe shortening for all four eyelids.

The patient was first seen in the ophthalmology clinic when she was five years old. She presented with red eye, was diagnosed with dendritic ulcer and managed accordingly. Since then, she has been followed up regularly. One year after her initial presentation, she was found to have bilateral cicatricial ectropion of the lower eyelids and diffuse superficial punctate keratopathy. She was treated with

vitamin A ointment and frequent lubrication. Her condition progressed into the development of exposure keratopathy with poor vision measuring 20/200 on both eyes.

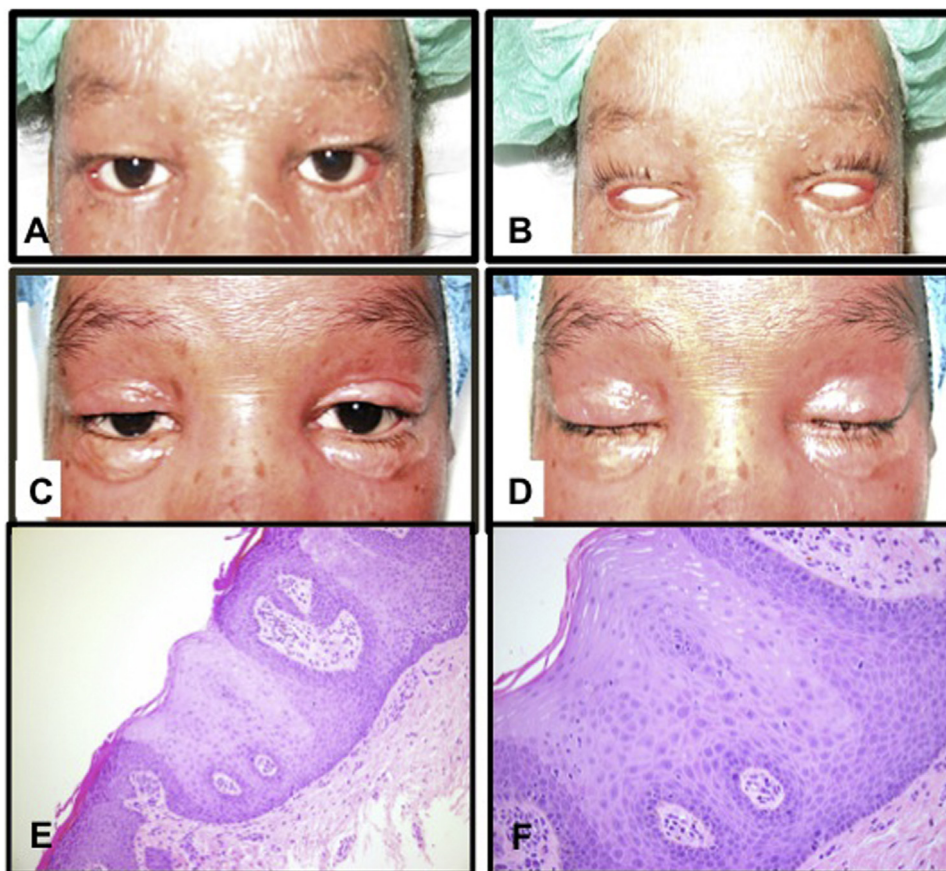
At the age of fourteen, she was referred for the first time to our oculoplastic service. She presented to us with burning sensation and decreased vision bilaterally. At that time, her visual acuity of the right eye was counting fingers at 6 feet and measured 20/200 on the left eye with no improvement using the pinhole. Her ophthalmic evaluation revealed severe bilateral lower and upper eyelids cicatricial ectropion (Fig. 1A and B). Slit lamp examination showed marked punctate erosions in both corneas with no signs of infection along with diffuse faint corneal scarring. The extra-ocular motility was full in both eyes and there was no afferent pupillary defect. Her management plan was to perform surgical repair of the cicatricial ectropion in all eyelids using MMG taken from the oral cavity.

The patient tolerated the surgery well and showed good healing postoperatively. She has been followed up for six years. Clinically, the graft has been viable without signs of infection and the patient had no further complaints (Fig. 1C and D).

A biopsy was taken from the MMG graft six year postoperatively after obtaining an informed consent from the patient. The biopsy showed squamous mucosa with acanthosis, keratinization and no signs of dysplasia or malignancy (Fig. 1E and F).

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**Fig. 1.** (A&B) The preoperative appearance of the bilateral lower lid cicatricial ectropion. (C&D) The clinical appearance, six years post-treatment with mucous membrane graft of both lower lids. (E) The histopathologic appearance of the acanthotic mucous membrane graft with evidence of metaplasia and keratinization (Original magnification x 100 Hematoxylin and Eosin). (F) Higher magnification of the metaplastic mucosa several years following the graft procedure. Note the absence of dysplasia. (Original magnification x 200 Hematoxylin and Eosin).

### 3. Discussion

Inherited Ichthyosis is defined as a heterogeneous group of disorders that are characterized by abnormal hyperkeratinization, subsequent scaling and loss of normal skin integrity. Depending on its association with other organs' abnormalities, ichthyosis is categorized into syndromic and non-syndromic forms. Abnormalities in other organs characterize the syndromic form such as in Netherton Syndrome and Sjogren-Larsson Syndrome, whereas the skin is the only expression of the disease in the non-syndromic form. Non-syndromic ichthyosis includes Autosomal Recessive Congenital Ichthyosis, Keratinopathic Ichthyosis, Ichthyosis Vulgaris and Recessive X-linked Ichthyosis [1,2].

Many ocular manifestations have been described in the literature to be associated with ichthyosis. One of the most common ocular complications of ichthyosis is cicatricial ectropion secondary to shortening of the anterior lamella [3]. The first reported ichthyosis-related ectropion was by Arnold in 1834. Following this, many cases were reported [5]. Most of the cases were found in patients with Autosomal Recessive Congenital Ichthyosis, in particular with lamellar and Harlequin clinical subtypes [4].

Ectropion can result in many medical and cosmetic consequences. Medical complications include exposure keratopathy and perforation, conjunctivitis and lagophthalmos. Patients also suffer from photophobia, pain, tearing and foreign body sensation. In addition, we can't underestimate its cosmetic psychological effects [6].

Cicatricial ectropion is generally managed medically and surgically. Medical treatment is conservative to eliminate exposure

side effects. Patients are often advised to use lubricants [7]. The definitive management is through surgical correction of the eyelids' malposition by the use of grafts. However, until present, surgical repair has been difficult and outcomes are still uncertain. Despite the huge variety of procedures done, no single procedure has been standardized [8]. In many occasions, it is hard to find healthy skin to be used as a graft in patients with Ichthyosis [6]. Mucus membrane on the other hand is thin and is not affected by the disease process. Using MMG was reported in 2011 in a patient who had bilateral cicatricial ectropion of both upper and lower lids. The MMG in that case was similarly taken from the oral cavity and the patient had improved symptoms with six months follow [9]. Here we report six years follow up for the clinical outcome using MMG, which is the longest follow up period reported confirming survival of the graft and the acceptable cosmetic appearance in addition to preserving the functionality of the eyelid. Histopathologically, the MMG showed evidence of acanthosis and mild metaplasia but no alarming signs of dysplasia or marked fibrosis and contracture. Thus we believe that MMG can be standardized in the management of these cases.

Our case has been prepared and reported in line with the SCARE criteria in: "The SCARE Statement: Consensus-based surgical case report guidelines". International Journal of Surgery 2016; 34:180-186. The authors further stress that they have no financial disclosures related to their recommendations [10].

In conclusion, Mucous Membrane Grafting is a promising option which can be offered for ichthyosis patients with cicatricial ectropion. A good outcome is expected using this technique due to sparing of the mucous membrane from being involved in the dis-

ease process as we have demonstrated by tissue diagnosis. Thus, we believe that MMG in such patients will result in the relief of the ectropion and prevention of its consequences along with good healing, and acceptable cosmetic appearance.

### Declaration statement

The authors have no financial or conflict of interest related to this work. An informed General Consent has been taken, which includes using patient's anonymous information.

### Author contribution

First author: Histopathology and manuscript review and corresponding author.

Second author: Review of chart, literature review and drafting of the case report.

Senior author: Surgical care of the patient and manuscript review.

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### Ethical approval

Case reports do not require approval, however the General informed consent includes patient's approval for use of relevant clinical and surgical information in an anonymous way.

### Consent

General informed consent has been taken.

### Conflicts of interest

The authors have no conflict of interest related to this case report.

### Guarantor

Dr. Hind Manaa Alkatan.

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