

Complex surgical treatment of Blepharophymosis syndrome (BPES)

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Purpose: to present blepharophymosis–ptosis-epicanthus inversus-telecanthus syndrome and its treatment options. BFES is an autosomal dominant condition linked to FOXL2 gene, approximately 50 % are sporadic.

Methods: retrospective case series of patients with BPES who underwent surgery at our hospital. Correction consists of correction of canthi and correction of ptosis. Traditionally it involves multiple steps; first medial canthoplasty to address blepharophimosis, epicanthus inversus and telecanthus in children of age 3 to 5 years, followed by correction of ptosis after approximately 1 year mostly by frontalis suspension or resection of LPS, Surgical treatment of blepharophimosis is very complex due to changed histological and anatomical properties of the orbit and its contents, we perform the treatment **on case per case basis** We have had very good results using Y-V technique for medial canthal correction and subtotal levator resection for correction of ptosis with removal of the fibrous plate whenever possible.

Results: We observed good cosmetic and functional results in all of our patients as evidenced by measurement of pre- and postoperative vertical intrapalpebral fissure height and MRD. Using levator resection we obtained better aesthetic outcome with more natural appearance compared to traditional frontalis suspension due to preserved palpebral fold. No intraoperative or postoperative complications were recorded. In some cases we noticed undercorrection of ptosis and scarring of medial canthus, which resolved by time.

Conclusions: Each patient with blepharophymosis syndrome should be approached individually as the expression of syndrome and hence patients' symptoms are variable. From our experience we prefer to use multi-step approach. Whenever possible we prefer to use levator resection for the correction of ptosis since it provides superior aesthetic and functional results.