

## **PENATALAKSANAAN BEDAH UNTUK SINDROMA BLEFAROFIMOSIS (PROSEDUR SATU TAHAP)**

### **Abstrak :**

*Objective: To report a one-step surgical management for blepharophimosis syndrome. Methods: A case report of 4 y.o boy with blepharophimosis syndrome. Results: A 4 y.o boy presented with common features of blepharophimosis syndrome (e.g. blepharophimosis, telecanthus, epicanthus inversus and bilateral ptosis). Examination revealed his VA were 2/60 RE and 5/60 LE, head turn to the right, distance of medial canthal and lateral canthal was 20 mm / 20 mm, FIP 0 mm / 1 mm, MRD 0 mm / 0 mm, MLD 0 mm / 0 mm on RE and LE respectively. Ocular motility and Bell's phenomenon were hard to be evaluated due to narrow eyelids fissure. We decided to perform a one-step reconstruction surgery on both eyes. First we correct epicanthus inversus and telecanthus with Mustarde procedure (combined of double Z and Y-V plasty), followed by ptosis correction with Crawford frontal-sling procedure using fresh fascia lata. One month later his FIP was 6 mm and 5 mm, MRD 2 mm and 1 mm, and MLD 4 mm on both eyes respectively, with slight lagophthalmos and positive bell's phenomenon. There were head turn to the right, nystagmus in RE when LE was closed, and decreased of RE macular reflex. We believe that these conditions due to undeveloped macula on RE. Conclusion: Blepharophimosis syndrome is a clinical entity which is characterized by presence of a small palpebral aperture in its overall dimensions. It is a congenital condition in which apart from blepharophimosis there is presence of telecanthus, epicanthus inversus and bilateral ptosis. This condition causes a great cosmetic blemish and hence its surgical treatment is most desirable.*

### **Keyword :**

*Keywords: blepharophimosis syndrome, telecanthus, epicanthus inversus, ptosis*

*Keywords: blepharophimosis syndrome, telecanthus, epicanthus inversus, ptosis  
blepharophimosis syndrome, telecanthus, epicanthus inversus, ptosis*