

눈꺼풀처짐을 수반한 눈구석벌어짐증

백봉수 · 지소영 · 최재일 · 석정훈 · 양완석

동강병원 성형외과

Telecanthus Associated with Blepharoptosis

Bong Soo Baik, M.D., So Young Ji, M.D., Jae Il Choi, M.D.,
Jeong Hoon Suhk, M.D., Wan Suk Yang, M.D.

Department of Plastic and Reconstructive Surgery, Dong
Kang General Hospital, Ulsan, Korea

Purpose: Blepharoptosis is often associated with telecanthus and the presence of epicanthal fold in telecanthus is one of unique features in Asian eyelids. The purpose of this article is to define telecanthus and pseudotelecanthus, and to determine optimal surgical procedure depending on classification of telecanthus.

Methods: Among 187 patients with blepharoptosis who had the advancement procedure of the Muller's musclelevator aponeurosis composite flap for ptosis, 55 patients underwent Flowers' split V-W plasty concomitantly with shortening the medial canthal tendon for correction of telecanthus from September 2003 to January 2011. Among them, 52 patients were followed up for 16 months. We newly defined telecanthus because Mustarde ratio is inaccurate to measure in certain cases and then made a definition of pseudotelecanthus. Besides, we also classified telecanthus into mild, moderate and severe types based on its severity.

Results: Telecanthus is defined when the ICD (inner canthal distance) is greater than 110% of normal ICD. Pseudotelecanthus is a telecanthus like a wide skin bridge formed between the eyes because of the epicanthal fold in the normal ICD. Flowers' split V-W plasty combined with shortening medial canthal tendon was very effective in mild and moderate telecanthus with almost invisible scar and no recurrence occurring. In severe types, however, it showed high incidence (28%) of incomplete correction of telecanthus.

Conclusion: New definition of telecanthus can be easily

applied to any case and we think the classification of telecanthus is useful to select an appropriate operative procedure. Split V-W plasty with shortening of medial canthal tendon is a very effective procedure in mild and moderate telecanthus. Besides, it is also effective in improving the treatment outcomes of ptosis in cases of blepharoptosis associated with telecanthus.

Key Words: Telecanthus, Blepharoptosis, Pseudotelecanthus

I. INTRODUCTION

A telecanthus is referred to as condition where the distance between the medial canthi is relatively larger compared to the distance between pupils, and it is formally defined as a Mustarde ratio (ICD/IPD) being greater than 0.55.¹ Here, the ICD (inner canthal distance) is the distance measured between the medial canthi, and the IPD (interpupillary distance) is the distance measured between the centers of the bilateral pupils. With the conversion of Mustarde ratio to a percentile value, the case in which ICD is greater than 55 percent of IPD is defined as telecanthus. A diagnosis of telecanthus is restrictively made, however, for cases in which the IPD is normal. If the increased ICD should be combined with the increased IPD, caused by excessive width of the nose, or increased interorbital width, the corresponding case would be diagnosed of hypertelorism or ocular hypertelorism.²

The problem of Mustarde ratio lies in the difficulty of making a diagnosis of telecanthus in some cases such as non-cooperative children, cases of nystagmus or strabismus and those in which the pupil is covered by the ptotic eyelid because the IPD can not be accurately measured. When an epicanthal fold is present, the ICD is referred to as the distance between the medial canthi, not the distance between the overlapping epicanthal folds which is IED (interepicanthal distance). When the ICD is normal and the IED/IPD is lower than 55 percent, a diagnosis of simple epicanthal fold can be made. In case in which the ICD is normal but the IED/IPD is

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Address Correspondence: Bong Soo Baik, M.D., Department of
Plastic & Reconstructive Surgery, Dong Kang General Hospital,
123-3, Taewha-dong, Jung-gu, Ulsan 681-320, Korea. Tel: 052)
241-1324/Fax: 052) 241-1324/E-mail: csy0203@han mail.net

greater than 55 percent due to wide epicanthal fold, however, a diagnosis of telecanthus cannot be made and no definite diagnosis is available at the present.

In this study, we introduce the method for easy making a diagnosis of telecanthus even in case in which the IPD can not be accurately measured. We have also coined a new term for case in which the ICD is normal but the IED is increased to the same extent as telecanthus. Furthermore, we have also classified telecanthus based on the severity of telecanthus and then described the optimal treatment modalities for each classification with our surgical treatment outcomes.

II. MATERIALS AND METHODS

Among 187 patients who underwent blepharoptosis surgery at our clinic between September 2003 and January 2011, 55 patients had combined diagnoses of blepharoptosis and telecanthus. Telecanthus and pseudotelecanthus were defined and telecanthus was classified based on its severity.

A. A novel definition of the telecanthus

The five standard measurements for the tissue around eyes include the inner canthal distance (ICD), outer canthal distance (OCD), interpupillary distance (IPD), interepicanthal distance (IED) and palpebral fissure length. Normally, the ICD is approximately 1/2 of the IPD and approximately 1/3 of OCD.³ Telecanthus is formally defined as Mustarde ratio (ICD/IPD) being greater than 0.55. It can be inferred that the ICD exceeding the IPD by 55 percent represents the addition of more than 5% of the IPD to the normal value of ICD. Because the IPD is twice greater than the ICD, any case in which more than 10% of the ICD is added to the normal value of ICD, based on the magnitude of ICD, is diagnosed as telecanthus. Consequently telecanthus can be referred to as a condition where the value of ICD is greater than its normal value by more than 110%.

B. The classification of telecanthus

Telecanthus can be classified as a pseudotelecanthus and a telecanthus. Then, based on the severity, a telecanthus can also be further divided into mild, moderate and severe types. The mild type is referred to as case in which the ICD is increased by 10~19 percent of the normal value; the moderate type is referred to as case in which the ICD is increased by 20~29 percent of the normal value; and the severe type is referred to as case in which the ICD is increased by more than 30%

compared with the normal value.

C. A pseudotelecanthus

When the ICD is normal and the IED is lesser than 110 percent of normal value of ICD, the corresponding case is diagnosed as a simple epicanthal fold.³ When the ICD is normal but the IED is greater than 110 percent of the normal value of ICD, however, it creates an illusion of telecanthus but a diagnosis of telecanthus can not be made. To differentiate this condition from a simple epicanthal fold, we can call 'pseudotelecanthus'. In most of the East Asian people who have a telecanthus, there is a concomitant presence of the epicanthal fold. To date, however, there has been no clear differentiation between simple epicanthal fold, pseudotelecanthus, telecanthus and telecanthus associated with epicanthal fold. Nevertheless, it can be helpful for selecting the treatment of choice to make a differentiation between them.

D. The determination of optimal surgical treatment depending on the classification of telecanthus

86.7% of Korean adults have epicanthal folds.⁴ A majority of patients with telecanthus also have epicanthal folds. For the pseudotelecanthus and the mild types of telecanthus, the methods which are frequently used to correct the simple epicanthal fold can also be used.⁵⁻⁷ However, we have used a split V-W plasty procedure of Flowers,⁸ a modification of Uchida method,⁹ for pseudotelecanthus and all types of telecanthus except one blepharophimosis syndrome. With this procedure, we also concurrently used a plication of the medial canthal tendon for shortening of the tendon in all types of telecanthus because of lengthening of the medial canthal tendon in telecanthus. For the case of blepharophimosis syndrome, telecanthus was corrected by a rectangular flap procedure of Mustarde¹ and transnasal wiring between medial canthal tendons.

E. Surgical technique of a split V-W plasty procedure of Flowers and shortening of medial canthal tendon

Marks are made on the bilateral palpebral skin where they are desired to place a new canthus and should be balanced on the skin surface between the eyes at the height of medial canthus. If the ICD is normal and the medial canthus is covered by the epicanthal fold, the mark should be placed on the anterior skin of the epicanthal fold at the projected site of the actual canthus. A triangular flap with a 60° angle and the same length (3~5 mm) of sides is designed around the mark. Here,

the tip of the flap is directed to the ipsilateral pupil and the proposed new canthal mark is placed on the line connecting between the tip of the flap and the midpoint of the base of the flap. In case in which the shortest distance between the new canthal mark and the margin of epicanthal fold is smaller than 5 mm, the location of new canthal mark is placed on the line between center and flap base. In other case, mark is placed on the base line of the flap. In any case, the tip of a triangular flap should not go over the margin of epicanthal fold. In case of pseudotelecanthus or mild and moderate telecanthus, a vertical line or a slightly bent line is drawn on or near the margin of epicanthal fold and the tips of W-character containing a 60° angle of central triangular flap are contacted to the vertical line. Thus, the triangular flap is placed between two triangles. Then, from the center of the vertical line to the actual canthus, a new line which the length is identical to the side of a triangular flap is drawn (Fig. 1, above, left). In severe telecanthus, there is a surplus amount of the skin for excision. In such a

manner that the vertical line should be slightly bent vertical line along the epicanthal fold, and the external length of W-character is drawn to be more longer to touch the bent line depending on severity of telecanthus. The tip of the central triangular flap can not reach this bent line (Fig. 1, Above, right). The length of the side of the triangular flap varies also depending on the severity of telecanthus. In case in which the length of the side of triangular flap exceeds 5 mm, however, scars can be visible on the skin and the newly formed canthus becomes too blunt. Because the epicanthal fold can be incompletely corrected in case in which it is smaller than 3 mm, a length of 3~5 (usually 4)mm would be appropriate. Following the incision based on surgical designs, the skin on two triangles which are located superior and inferior to the flap is excised and then the triangular flap is left between the two raw triangular surfaces. Another two triangular flaps are developed between the center of the vertical line and actual canthus, and these three triangular flaps are elevated. After removal of soft tissue



Fig. 1. (Above, left) Preoperative design of Flowers' split V-W plasty in mild and moderate telecanthus. The tips of W are contacted to the vertical line. (Above, right) Preoperative design of modified Flowers' split V-W plasty in severe telecanthus. To excise the surplus skin, two external tips of W are contacted to the bent vertical line but the tip of central triangular flap is not reached. (Below, left) Following incision on surgical designs, the skin on two triangles which are located superior and inferior to the triangular flap is excised and subcutaneous soft tissue is removed to expose the medial canthal tendon. The medial canthal tendon is plicated to shorten the length of tendon. (Below, right) Immediate postoperative findings after advancement of triangular flap toward the medial canthus and skin sutures.

including some part of orbicularis oculi muscle beneath the three triangular flaps, the medial canthal tendon is exposed.

The most lateral part of this tendon is sutured using a 6-0 prolene twice and the same needle is passed through the most medial part of the tendon or nasal periosteum. After ligation, the length of medial canthal tendon is shortened by approximately half (Fig. 1, Below, left).

The knot of prolene is covered with the adjacent soft tissue to prevent exposure. The subcutaneous tissue at the tip of each triangular flap is sutured using a 6-0 vicryl at the site where the triangle should be placed. Then, the skin is sutured using a 7-0 nylon (Fig. 1, Below, right).

III. RESULTS

Among 187 patients who had blepharoptosis correction with advancement of Muller's muscle and levator aponeurosis as a composite flap, 55 patients (29.4%) concurrently underwent surgery for telecanthus. Total blepharoptosis patients consisted of 91 mild ptosis, 72 moderate ptosis and 24 severe ptosis patients. Six of 91 mild ptosis (6.5%), 29 of 72 moderate ptosis (40.2%) and 20 of 24 severe ptosis patients (83.3%) had surgery for telecanthus. The more severe degree of ptosis, the higher frequent was incidence of telecanthus. Besides, 55 cases of telecanthus consisted of 35 male (63.6%) and 20 female (36.4%) patients with a male predilection.

In regard to the age distribution, there were one

patient aged between 0 and 10 years, 28 patients aged between 11 and 20 years, 20 patients aged between 21 and 30 years, 2 patients aged between 31 and 40 years and 4 patients aged 41 years or older. These results indicated that patients aged between 11 and 30 years accounted for 89.1%. Of total patients who were surgically treated, there were 5 cases of the pseudotelecanthus, 15 cases of the mild type, 24 cases of the moderate type and 11 cases of the severe type. As the surgical methods, surgery was performed to correct telecanthus and this was followed by the surgical correction of ptosis. Of 55 patients with telecanthus, one with blepharophimosis syndrome underwent a rectangular flap technique of Mustarde and in the remaining 54 cases, a split V-W plasty procedure of Flowers was concomitantly done with the folding of medial epicanthal tendon. Of 55 patients who underwent surgery for telecanthus, 52 were followed up during a mean period of 12 months (range: six months-four years).

Postoperative results of pseudotelecanthus, mild and moderate type of telecanthus showed good treatment outcomes with disappearance of epicanthal fold and normal ICD. In the severe type, however, 3 of 11 cases had incomplete correction of telecanthus which remained as a mild type of telecanthus. As the other postoperative complications, there was one case of hematoma which was resorbed spontaneously. Mild scar tissue was found on the medial palpebral skin in 4 patients, but there was no patient who concerned about it (Table I, Fig. 2-6).

Table I. Complications of the Split V-W Plasty with Plication of the Medial Canthal Tendon for Telecanthus

Complications	Pseudotelecanthus N : 5	Mild form N : 15	Moderate form N : 24	Severe form N : 11
Incomplete correction	–	–	–	3 (28%)
Visible scar	–	–	2 (8%)	3 (28%)
Hematoma	–	–	–	1 (10%)



Fig. 2. Operative result of pseudotelecanthus with severe blepharoptosis. (Left) Preoperative view. OCD: 92 mm. Calculated ICD: 31 mm, IED: 37 mm. (Right) 34 months postoperative view. ICD: 32 mm.



Fig. 3. Operative result of mild telecanthus with left mild blepharoptosis. (Left) Preoperative view. IPD: 66 mm. Ideal ICD: 33 mm, (Real) ICD: 38 mm. (Right) 38 months postoperative view. ICD: 34 mm.



Fig. 4. Operative result of moderate telecanthus with right moderate blepharoptosis. (Left) Preoperative view. IPD: 66 mm. ICD: 41 mm. (Right) 26 months postoperative view. ICD: 35 mm.



Fig. 5. Operative result of severe telecanthus with moderate blepharoptosis. (Left) Preoperative view. IPD: 68 mm. ICD: 45 mm. (Right) 4 months postoperative view. ICD: 36 mm.



Fig. 6. Operative result of severe telecanthus with moderate (Right) to severe blepharoptosis (Left). (Left) Preoperative view. IPD: 67 mm. ICD: 45 mm. (Right) 37 months postoperative view. ICD: 40 mm. Mild scar near the medial canthus and undercorrection of telecanthus were shown.

However the rectangular flap technique of Mustarde for a blepharophimosis syndrome case showed remarkable scars.

IV. DISCUSSION

Mustarde¹ first coined a term 'telecanthus' in case in which the IPD was normal and the distance between the medial canthi was abnormally increased. According to him, a telecanthus was defined as the ratio of inner canthal distance to the interpupillary distance (ICD/IPD) exceeding 0.55.

Because of difficulty and inaccuracy to measure the IPD in some cases, we defined a telecanthus as the increased ICD being greater than 110 percent of its normal value based on the ICD itself. If IPD can not be measured, the normal value of ICD corresponds to approximately 1/3 of OCD. Accordingly, the normal value of ICD can be measured in any case. According to Hwang et al.,⁴ the ratio of ICD/IPD is 1:1.89 in Korean adults and its percentile value is smaller than 53%. It can therefore be inferred that a diagnosis of telecanthus based on Mustarde ratio of ICD/IPD exceeding 55% can also be applicable to Korean people.

The epicanthal fold was defined, by Duke-Elder,¹⁰ as "a semilunar-fold of skin running downwards at the side of the nose with its concavity directed to the inner canthus." According to the anatomical locations where the epicanthal fold is developed, he classified the epicanthal fold into four types: epicanthus supraciliaris, epicanthus palpebralis, epicanthus tarsalis and epicanthus

inversus. From a surgical perspective, however, Mustarde³ classified the epicanthal fold into the following:

1. A simple epicanthal fold
2. An epicanthal fold associated with ptosis
3. An epicanthal fold-with or without ptosis-associated with telecanthus

According to Mustarde's classification system, an epicanthal fold can be associated with telecanthus. Although these two entities are different diagnoses, the surgical procedure for treatment has not been clearly differentiated between the simple epicanthal fold and the telecanthus associated with epicanthal fold, and most previous reports were described under the title of epicanthal fold. Accordingly, reports about telecanthus have been made in a limited scope.

The incidence of telecanthus has been reported to be relatively high in patients with cleft lip and palate.³ But there was no reported incidence of telecanthus in blepharoptosis patients up to present. In our series, the incidence of telecanthus in blepharoptosis patients was 29%, being relatively high. Because this incidence was reported from patients who were surgically treated for ptosis and telecanthus, it can not be a good data. There was also higher frequency of telecanthus following the more severe degree of blepharoptosis.

We classified a telecanthus into pseudotelecanthus, mild, moderate and severe type of telecanthus depending on its severity. It would be rational to apply the surgical treatment modality to each type. In regard to the treatment, for a pseudotelecanthus or the mild type of telecanthus, such procedures as medial epicanthal plasty using skin

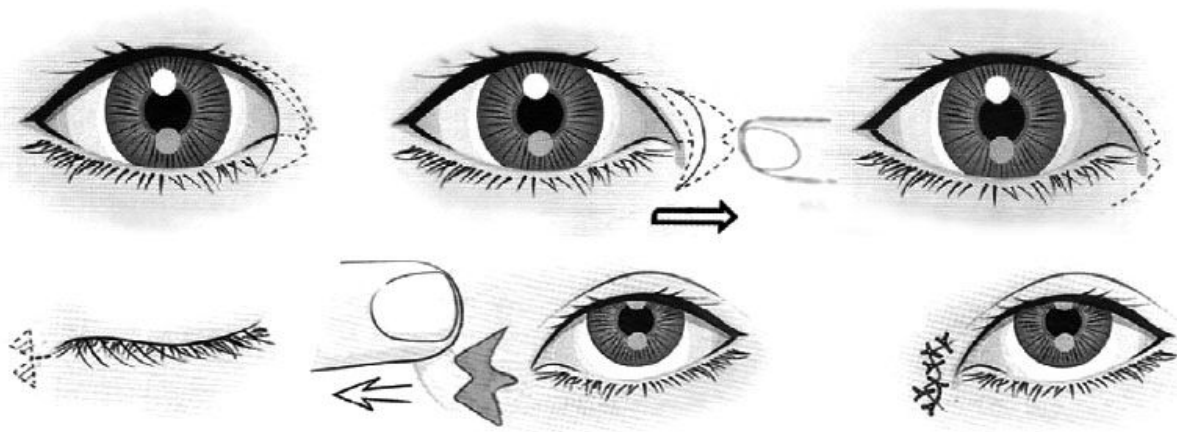


Fig. 7. (Above) Uchida's triangular flap method. (Above, left) Preoperative design. (Above, center) Dotted lines are preoperative designs and solid line is epicanthal fold line. (Above, right) Post operative scar line. (Below) Flowers' split V-W plasty methods. (Below, left) Preoperative design of split V-W plasty procedure. The area of dashed line will be excised. (Below, center) The wound after incision and medial traction. (Below, right) State of closure of the wound with 7-0 Nylon suture. (Quoted from *Cosmetic & Reconstructive Oculoplastic surgery*, 3rd ed, Seoul, Gunja, 2009, p. 394)

redraping method,⁵ W-epicanthoplasty⁶ or Z-epicanthoplasty⁷ can be selectively used as done for a simple epicanthal fold. Uchida⁹ reported the method where the redundant skin of the epicanthal fold was resected in a triangular shape and the linear scar contracture was prevented with the advancement of a small-sized triangular flap to the direction of epicanthal fold (Fig. 7).

Besides, Flowers reported a modified method of Uchida, a split V-W plasty technique⁸ (Fig. 7). The Uchida method and Flower's modification were basically the procedure for epicanthal fold and they did not describe the shortening procedure of the medial canthal tendon.

In all types of telecanthus, we performed a split V-W plasty technique of Flowers in combination with shortening of medial canthal tendon but there was high incidence of incomplete correction in severe type of telecanthus. In the severe type, we recommend a method of Uchida⁹ where the redundant skin on the medial palpebral area can be more resectable than Flowers' modification as far as scars and tension of suture line were acceptable (Fig. 7, above). We also selected the methods where the medial canthal tendon was shortened as maximally as possible in severe type. In one case, in which the medial canthal tendon was very long because of severe blepharophimosis syndrome, a transnasal wiring was used. Mustarde's four flap technique caused remarkable scar formation on the medial palpebral skin, so its alternative one has been sought after.¹¹ The Mustarde technique is only considered in such limited cases as severe blepharophimosis syndrome.

Converse et al.² defined hypertelorism as an increased distance between the medial orbital walls (interorbital distance) including the increased IPD, with abnormally large ICD and OCD. Besides, case in which only the ICD was increased but the IPD and the distance between the orbits were normal was defined as a pseudohypertelorism rather than a telecanthus. Waardenburg's syndrome, cases of the increased ICD due to the naso-orbitoethmoidal fracture, blepharophimosis syndrome and various types of wide epicanthal folds were included into pseudohypertelorism. We would suggest a diagnosis of simple epicanthal fold in case in which both the IPD and the ICD were normal and IED was smaller than 110% of the normal ICD. We also would suggest a new diagnosis of a pseudotelecanthus in case in which the IED was greater than 110% of the normal ICD in the

condition of normal ICD and IPD.

Besides, because most of the Asian people have concomitant presence of epicanthal folds if they have telecanthus, we would suggest a diagnosis as 'telecanthus' rather than a diagnosis of 'epicanthal fold associated with telecanthus'.³

V. CONCLUSION

Telecanthus is often associated with blepharoptosis. We suggested new definition of pseudotelecanthus and telecanthus. The telecanthus can be classified into mild, moderate, and severe type based on its severity. The split V-W plasty technique of Flowers in combination with shortening of the medial canthal tendon was very good procedure with a minimal complication rate for the treatment of the mild and moderate type of telecanthus but there was high incidence (28%) of incomplete correction of telecanthus for the severe type of telecanthus. Concurrent correction of the telecanthus helped clearly to improve the operative results of blepharoptosis in case of telecanthus associated with ptosis.

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