

Evaluation of the complex treatment for congenital blepharoptosis

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OBJECTIVE: To evaluate the results of treatment of congenital blepharoptosis (CBP) using selected surgical methods; and to evaluate concomitant visual system disorders.

METHODS: Between 2001 and 2010, 52 children with CBP underwent surgical correction of CBP using the modified method of Mustarde, the original Mustarde method or frontal suspension at the Department of Plastic Surgery, Medical University of Lodz (Poland). Based on the results of ophthalmic and orthoptic examination, and standard measurements, postoperative differences in the position and symmetry of the upper eyelids, complications, and visual system abnormalities were analyzed.

RESULTS: Very good results were obtained in all patients with mild unilateral CBP. The results in patients with moderate and severe unilateral CBP, and in bilateral anomaly after correction using Mustarde's method or the modified Mustarde's method, were also very good. Complications included lagophthalmos (15.4%) and undercorrection (3.8%). Visual system disorders, mainly amblyopia, resulting from strabismus, astigmatism, anisometropia and CBP, were observed in 88.5% of patients

CONCLUSIONS: Complex ophthalmic examination and measurements in individuals with CBP enable correct diagnosis, selection of appropriate treatment method and timing of surgical intervention. Postoperative results in patients with CBP supported the efficacy of the methods that shortened the levator palpebrae superioris. Supplementing with Mustarde's modified method contributed to an increase in the number of favourable postoperative results.

Key Words: *Congenital blepharoptosis; Ophthalmologic abnormalities; Surgical treatment*

The first textbook of ophthalmology and ophthalmic surgery published in 1583 by Georg Bartish (1) includes references to blepharoptosis and some illustrations of simple methods facilitating elevation of the upper eyelid; however, the author did not mention the congenital background of this anomaly. Congenital blepharoptosis (CBP), a common developmental disorder of the palpebral apparatus, occurs in 7.9 per 100,000 individuals (2). Although the cause of CBP remains unknown, most authors who support the myogenic theory state that dystrophy or dysgenesis of the levator palpebrae superioris (LPS) leads to abnormal performance (3,4). Maldevelopment of the LPS occurs in >50% of CBP cases. Autosomal-dominant forms of isolated anomaly are associated with the *PTOS1*, *PTOS2* and *ZFH-4* genes (5). In other cases, CBP is related to congenital syndromes with myogenic origin, such as blepharophimosis, generalized extraocular muscle fibrosis and myasthenia, or those of neurogenic etiology such as Marcus Gunn syndrome, oculomotor nerve palsy and Horner syndrome (6,7).

CBP is often associated with numerous visual system disorders, mainly amblyopia, strabismus, anisometropia or astigmatism, which should be well known both to ophthalmologists and plastic surgeons who eventually decide on surgical intervention (8,9). The treatment requires clinical analysis to select the appropriate operative method.

L'évaluation du traitement complexe de la blépharoptose congénitale

OBJECTIF : Évaluer les résultats du traitement de la blépharoptose congénitale (BPC) à l'aide de certaines méthodes chirurgicales et évaluer les troubles du système visuel s'y associant.

MÉTHODOLOGIE : Entre 2001 et 2010, 52 enfants ayant une BPC ont subi une correction chirurgicale au moyen de la méthode modifiée de Mustarde, de la méthode originale de Mustarde ou de la suspension aux muscles frontaux au département de chirurgie plastique de l'université médicale de Lodz, en Pologne. D'après les résultats de l'examen ophthalmique et orthoptique et les mesures standards, les chercheurs ont analysé les différences postopératoires dans la position et la symétrie des paupières supérieures, les complications et les anomalies du système visuel.

RÉSULTATS : Tous les patients ayant une BPC unilatérale bénigne ont obtenu de très bons résultats. Ceux dont la BPC unilatérale était modérée ou grave ou dont l'anomalie était bilatérale ont également obtenu de très bons résultats après correction par la méthode de Mustarde classique ou modifiée. La lagophthalmie (15,4 %) et la sous-correction (3,8 %) faisaient partie des complications. Les chercheurs ont observé des troubles du système visuel chez 88,5 % des patients, notamment l'amblyopie, causés par un strabisme, un astigmatisme, une anisométrie ou une BPC.

CONCLUSIONS : Chez les enfants ayant une BPC, un examen ophthalmique approfondi et des mesures favorisaient un bon diagnostic, le choix de la méthode thérapeutique appropriée et la sélection du bon moment pour effectuer l'intervention chirurgicale. Les résultats postopératoires chez les patients ayant une BPC corroboraient l'efficacité des méthodes visant à raccourcir le muscle releveur de la paupière supérieure. L'ajout de la méthode modifiée de Mustarde contribuait à accroître le nombre de résultats postopératoires favorables.

CBP surgery has been a topic of interest for many authors; however, despite a variety of advocated procedures for its correction, shortening of the levator muscle complex remains the best option to achieve satisfactory functional and aesthetic postoperative results (10,11).

METHODS

A group of 52 CBP patients (32 male, 20 female) between two and seven years of age underwent ophthalmologic and orthoptic examination and CBP surgery at the Department of Plastic, Reconstructive and Aesthetic Surgery of the Medical University of Lodz, Poland. Postoperative follow-up examinations one year later were performed at the Plastic Surgery Outpatient Department, and Binocular Vision Pathophysiology and Strabismus Department, Medical University of Lodz, during the period from 2001 to 2010.

Ophthalmologic examination included family history, visual acuity, refraction under full cycloplegia, degree of amblyopia, angle of deviation (measured using prism and alternate cover test – PACT, PCT – for distance and at near, using major amblyoscope), sensory status (evaluated using the Titmus stereo test, TNO test, Bogolini lenses), ocular movement measured using prism and alternate cover test (in nine positions by synoptophore and by Hess chart), compensatory face turn

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TABLE 1
Results of congenital blepharoptosis (CBP) surgery in patients with unilateral CBP

Preoperative degree of ptosis	Patients, n	Difference in palpebral margin position of the two upper eyelids (normal and corrected) in primary gaze following CBP surgery, mm		
		0–1	>1 to ≤2	2–4
Mustarde modified method				
Mild	2	2 (100)	–	–
Moderate	7	6 (85.7)	1 (14.3)	–
Severe	7	5 (71.4)	1 (14.3)	–
Total	16 (100)	13 (81.25)	2 (12.5)	1 (6.25)
Mustarde method				
Mild	4	4 (100)	–	–
Moderate	14	12 (85.7)	2 (14.3)	–
Severe	9	7 (77.8)	1 (11.1)	1 (11.1)
Total	27 (100)	23 (85.2)	3 (11.1)	1 (3.7)

Data presented as n (%) unless otherwise indicated

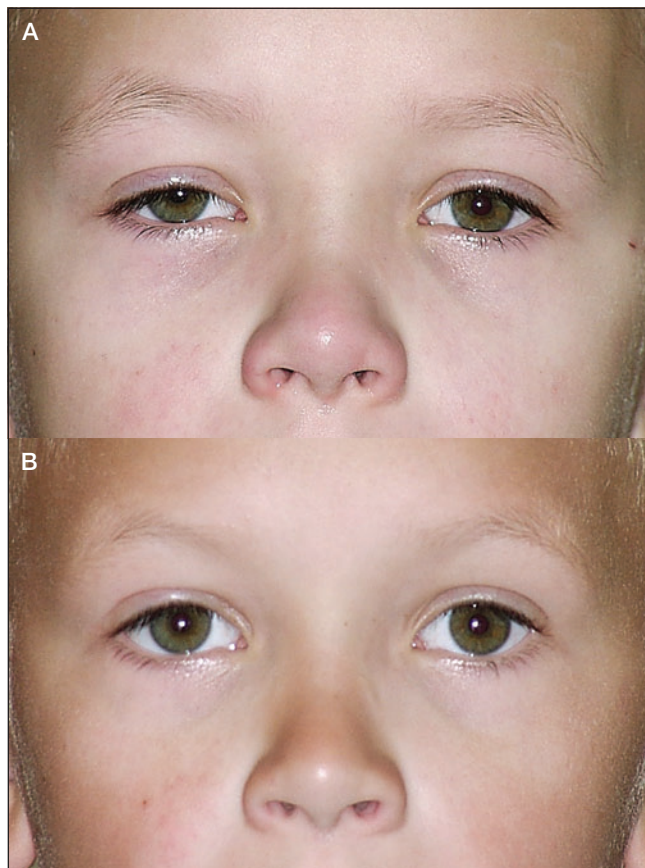


Figure 1 A Five-year-old boy with moderate unilateral congenital blepharoptosis. B One year after correction using Mustarde's modified method

and head tilt. The following parameters were included in the analysis: degree of amblyopia before surgery and postoperatively; compensatory head posture; change in astigmatism; presence of anisometropia; and presence of strabismus, especially hypotropia. Preoperative examination was performed in every case at the Department of Plastic Surgery, Medical University of Lodz, and included:

- LPS function (Beard's scale);
- Degree of ptosis (regarded as mild if eyelid dropped ≤2 mm, moderate if it dropped >2 mm but <4 mm, and severe if it dropped ≥4 mm);
- Heights of palpebral fissures and eyelid folds;
- Bell's phenomenon;
- Lagophthalmos; and
- Compensatory head posturing or brow(s) elevation

All patients were photographed while looking straight ahead, up and down. The degree of ptosis, taking into consideration uni- and bilaterality, determined the individual choice of surgical technique (for every corrected patient).

All 52 patients with myogenic CBP (43 unilateral; nine bilateral) were operated on: 18 cases using Mustarde's method; 30 using the modified Mustarde's method; and in four using frontal suspension (1a, b, 2a, b, 3a, b). Five children were <4 years of age.

Mustarde's method (split-level lid resection) consists of skin and orbicularis muscle resection at the upper level, and resection of 4 mm to 5 mm of tarsal plate with conjunctiva (subtotal resection) at the lower level. Mustarde advocated peripheral shortening of the levator complex according to Rycroft's rule (4 mm of LPS for every 1 mm of ptosis); this method was used in 18 cases. To minimize the range of lagophthalmos after Mustarde's technique, this method was modified and used a limited resection: 3 mm of the tarsus and 3 mm of LPS for every 1 mm of ptosis.

Frontalis muscle suspension with fascia lata sling was used in patients with severe CBP with no detectable LPS function.

The following measurements were re-examined and reassessed postoperatively, and compared with preoperative parameters:

- The position of the upper eyelid margin in relation to the cornea, with the pupil in primary position; in unilateral CBP difference in the height of the two palpebral fissures was considered to be the extent of ptosis, estimated both visually and from photographs; and
- Lagophthalmos

RESULTS

In patients with unilateral CBP, the authors defined a very good result when the postoperative difference of the position of the upper eyelids (normal and corrected) in primary gaze was 0 mm to 1 mm. Taking into consideration the pre- and postoperative palpebral positions in primary gaze, they obtained very good final results in all cases of unilateral mild CBP and in 85.7% of moderate CBP. In severe cases of CBP, very good results were confirmed in 71.4% of cases corrected by Mustarde's method and in 77.8 % of patients treated using Mustarde's modified method (Table 1).

In patients with bilateral CBP, a very good result was defined when the difference in the postoperative position of the upper lids was above the preoperative (in primary position) position: 1 mm to 2 mm in mild ptosis: 2 mm to 3 mm in moderate ptosis; and 3 mm to 5 mm in severe ptosis. Considering these criteria, very good results with symmetric eyelid position(s) were found after bilateral operation with LPS shortening and also after frontalis suspension in patients with moderate CBP (n=2) and with severe CBP (n=5).

There were no early postoperative complications (observed from the day of surgery up to one week). Late complications (from one week to one year) were noted in 19.2% of the examined group: in eight (15.4%) patients, lagophthalmos was present after correction using

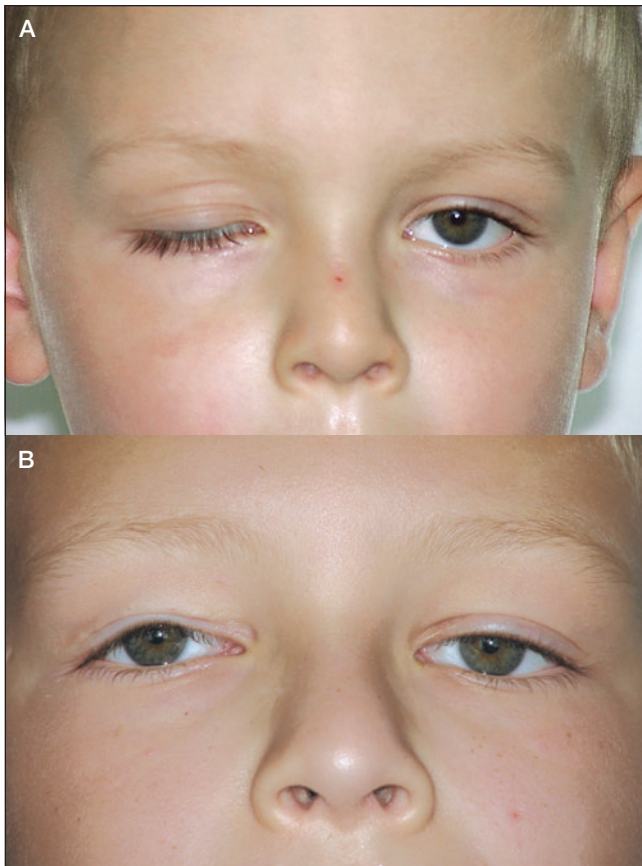


Figure 2) A Four-year-old boy with severe unilateral congenital blepharoptosis. **B** Four years after correction using Mustarde's modified method



Figure 3) A Seven-year-old patient with severe bilateral congenital blepharoptosis. **B** Two years after correction using Mustarde's modified method

Mustarde's modified method (n=3) and by Mustarde's method (n=5); none of whom exhibited incomplete lid closure before surgery. The range of lagophthalmos oscillated between 1 mm and 3 mm: in a single case with moderate CBP with 8 mm of LPS function; and in seven with severe ptosis with 6 mm of LPS function. In three patients operated on using Mustarde's modified method, lagophthalmos did not exceed 1 mm. However, in four severely affected cases, correction using the Mustarde method amounted up to 3 mm. The treatment in all of these cases included applications of bland ointments and artificial tears. Undercorrection (1 mm to 1.5 mm) was observed in two children with severe CBP; however, their parents did not agree to reoperation(s).

Ophthalmic examination and orthoptic evaluation revealed ophthalmologic abnormalities in 46 (88.5%) patients. In 23 (44.2%) cases, it was amblyopia, and in nine (19.5%) after blepharoplasty was diagnosed. Coexistence of strabismus in nine (39.1%) cases, anisometropia in five (21.7%), astigmatism in six (26.1%) and CBP in three (13.1%) children with amblyopia was observed. Compensatory head posture was confirmed in 18 (39.1%) patients, who presented with eight cases of mild or moderate amblyopia and had peripheral fusion in most of them. Astigmatism was confirmed in 32 (69.6%) patients, which was diagnosed in 10 (21.7%) cases preoperatively and observed in 22 (47.8%) after CBP surgery. Anisometropia was noted in five (10.8%) patients. In two cases, it was present pre- and postoperatively and, in the remaining cases, it appeared after surgery. In the group with confirmed strabismus, 14 (30.4%) children presented the manifest (horizontal and vertical) and 12 (26.1%) – the latent form. Strabismus was detected in nine (34.6%) cases preoperatively and in 17 (65.4%) postoperatively; thus, the diagnosis in eight children was made after surgery. This was due to the fact that performing a complete ophthalmological and orthoptic examination demands full

cooperation from the patient, which is very difficult in young children. This explains why strabismus was not detected preoperatively in eight cases, but only confirmed when cooperation with these young patients improved. Exophoria at near was observed in five cases and prevailed over other latent forms.

DISCUSSION

Similar to previous reports, we agree that the severity of CBP and LPS (dis)function, together with the presence of visual system abnormalities, as well as the age of the affected patients, should influence complex treatment based on clinical analysis, ophthalmologic and orthoptic examination, and standard measurements (12). Precise measurements can usually be performed in children between four and five years of age. Similar to others, we also recommend surgery for a ptotic eyelid before school age to avoid the consequences of premature correction; most of the cases in the present study underwent CBP surgery between four and seven years of age. However, in patients with a threatened pupil, early operation should be considered to avoid stimulus deprivation amblyopia. Accordingly, surgical correction of CBP was performed in five children <4 years of age in our group (8,13). Undoubtedly, LPS shortening (when LPS function is >3 mm to 5 mm) is the preferred method for CBP correction. The extent of LPS shortening depends on the degree of ptosis, and oscillates between 8 mm in mild CBP and >20 mm in severe cases, which alternatively, can undergo frontal suspension when LPS function is poor to absent. Fascia lata is the best material to suspend lids because of its biocompatibility (7,10,14,15). Four patients in our study underwent the bilateral suspension procedure and, in all cases, good postoperative results were observed. The majority of patients from the present series with poor (but >5 mm) LPS action underwent surgery with complex shortening using Mustarde's or Mustarde's modified method. Retrospective analysis of postoperative results proved

comparable efficacy of both techniques. Modifying Mustarde's technique enables functional and aesthetic improvement in CBP cases; hence, widens the spectrum of surgical corrective measures.

As the most common late complication, lagophthalmos of up to 3 mm was present in patients who underwent correction using an LPS resection method according to Rycroft's rule, and with subtotal resection of the tarsal plate. In patients with a similar severity of CBP and 6 mm of LPS function, lagophthalmos was confirmed after the operation using Mustarde's modified method and did not exceed 1 mm. Similar to other authors, we also confirm the association of lagophthalmos with the range of resected LPS and CBP severity (7,11,15). Our findings also indicate a correlation between postoperative incomplete lid closure and the amount of resected tarsal plate. This corresponds to a small number and the lower range of incomplete lid closure in children from our study who underwent correction using Mustarde's modified method. Some authors explain the presence of postoperative lagophthalmos by incomplete freeing of the septum orbitale, or sutures incorporating the septum and LPS aponeurosis, resulting in a 'hangup' with eyelid closure (16,17). Children's eyes are usually tolerant to drying and, in our group, cases with lagophthalmos corneas adapted with appropriate treatment.

Although amblyopia was the most frequently observed visual disorder in the study group, it was rarely caused by isolated CBP. However, its ratio increased if strabismus, astigmatism or anisometropia coexisted. Postoperative amblyopia in five (19.5%) cases in our group may have been a result of CBP surgery performed too early before the age of four years. If amblyopia is a consequence of CBP, then a decision of ptotic eyelid surgery should be made without delay. These observations are similar to the results reported in other studies (8,18).

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We are consistent with other authors who reported that compensatory head posture does not exclude the presence of amblyopia and may allow the maintenance of peripheral fusion (19). Ptotic eyelid pressure probably leads to corneal astigmatism, which was observed in 21.7% of our group before surgical correction, which changes the position of the operated lid and, hence, may be also responsible for postoperative astigmatism diagnosed in 47.8% of the presented cases. Astigmatism often requires refractive correction and amblyopia therapy is necessary before surgery (20).

We agree with authors who indicate that postoperative amblyopia may be influenced by anisometropia as a result of premature surgery. Pleoptic treatment occurred in 9.6% of patients in the present study, in whom anisometropia resulted in postoperative amblyopia (21). Strabismus concomitant with CBP was noted in 26.9% of patients in the study by Thapa (22). We confirmed the incidence of manifest strabismus in 30.4% (horizontal and vertical) of patients and latent strabismus in 26.1%. Exophoria at near predominated over other latent forms.

Various ophthalmological disabilities were confirmed in many children in the present study. The incidence of these disabilities, in addition to the frequencies reported by other authors, emphasize the importance of early evaluation and, if needed, prophylactic treatment of CBP patients. Thus, complete ophthalmic and orthoptic examination, together with standard measurements, should become an integral part of the complex treatment in individuals with CBP.

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