Dandy Walker Syndrome with Tessier 7 Cleft-a Rare Case Report and a Surgical Note on the Use of Vermilion Flap and Lazy W-Plasty

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Received: 6 May 2011 / Accepted: 26 September 2011 / Published online: 20 October 2011
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Abstract The Dandy-Walker syndrome is a malformation of the brain that involves the mal-development of the cerebellum, associated with a cystic enlargement of this area, and frequently hydrocephalus. This malformation occurs in ~1 in 30,000 babies. It is seen mostly in females. Developmental anomalies like cleft lip, cleft palate, and cardiac malformation, orthopaedic and urinary structural abnormalities may also occur in 30% of the individuals. We report a case of Dandy Walker syndrome with Tessier 7 facial cleft with paramedian cleft palate in a 6 month old child. Surgical methods used to correct this anomaly include commissuroplasty, myoplasty of the orbicularis oris, and closure of the cleft cheek. Authors report a vermilion square flap technique that combines a lower lip mucocutaneous vermilion border flap with a lazy W-plasty to ensure a natural commissure and skin closure.

Keywords Dandy-Walker syndrome (DWS) · Tessier 7 malformation

Introduction

Dandy-Walker syndrome is a congenital brain malformation involving the cerebellum and the fluid filled spaces around it. A key feature of this syndrome is the partial or even complete absence of the part of the brain located between the two cerebellar hemispheres. This syndrome is a genetically sporadic disorder that occurs one in every 30,000 live births with a slight female preponderance [1].

It was first described in 1914 by W. Dandy and K. Blackfan, later designated as Dandy Walker syndrome in 1954 by C. Benda [2].

Dandy-Walker syndrome is frequently associated with other intracranial anomalies such as agenesis of the corpus callosum, holoprosencephaly, occipital encephaloceles and ocular abnormalities. Extra-cranial abnormalities include polycystic kidneys, cardiovascular defects, polydactyly and cleft palate.

The purpose of this article is to delineate the importance of commissuroplasty with a vermilion square flap from the lower lip, and closure of the oblique facial cleft with a lazy W-plasty as an effective surgical procedure in order to obtain good clinical results.

Patient Report

A 6 months old boy was brought to the Department of Oral Maxillo Facial Surgery with a complaint of a deformed face and a wide mouth since birth. His perinatal history revealed that he was diagnosed with Dandy-Walker syndrome along with congenital heart disease.

Extra oral examination revealed gross facial asymmetry with a lateral facial cleft on the right side, extending from the commissure (which was absent) up to the anterior border of masseter. A linear depression extending from the cutaneous component of the cleft to the zygomatic arch demonstrated a subcutaneous component of the cleft with disruption of the underlying muscles. The right ear appeared smaller than the left ear. An epicanthal fold was seen on the right eye, while the left was normal. The lip on the unaffected side presented with good morphology and satisfactory competence (Photograph 1). Intraoral examination revealed a paramedian cleft of the soft palate.

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Computerised Tomography of the facial skeleton confirmed the absence of bony cleft of zygomatic bone or maxilla.

Management of this deformity was planned in two stages. Stage one procedure involved correction of macrostomia using vermilion rotational flap with modified W-plasty for the closure of oblique facial cleft. Stage two procedure surgical correction of the cleft palate.

Operative Technique

The steps followed in surgical correction were creation of straight-line closure of the mucosa, red-lip commissural flap rotated from the lower lip, modiolus muscle reconstruction and use of lazy W-plasty flap for cutaneous closure.

The midpoint of the upper lip was determined between the peaks of Cupid's bow. The length between the midpoint and the lateral end of the non-cleft side were transposed to the cleft side. A square mucocutaneous flap involving the vermilion border with its pedicle on the lower lip was designed to avoid a suture scar at the commissural point that may contract and deform the shape of the oral commissure (Photograph 1). A lazy W-plasty was planned to close the skin of the cleft cheek.

After injecting 2% xylocaine with adrenaline solution (1:2, 00,000), incisions were made and the excess tissue was excised. The skin and mucosa were dissected to identify the orbicularis oris muscle, thus exposing the muscle bundles which form the modiolus. To achieve a natural commissural shape the muscle bundles were overlapped, the upper bundle was placed anteriorly and the lower one placed posteriorly. These upper and lower muscles bundles were sutured with vicryl 4-0 sutures (Fig. 1). In addition, the muscle bundles remaining at the cleft site were sutured in a side-to-side fashion to fill the defect at the cheek (Fig. 2). After approximation of the

Photograph 1 intra operative photograph showing surgical markings

Fig. 1 figure showing the overlapping of the orbicularis oris muscle achieved during closure

Fig. 2 side to side suturing of the muscle bundles

Photograph 2 immediate post operative photograph showing closure of the skin

muscles, the skin was closed using lazy W-plasty technique (Photograph 2).

Discussion

Dandy-Walker syndrome is characterised by agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the

Photograph 3 post operative photograph after suture removal

fourth ventricle, and enlargement of the posterior fossa [3, 4]. Affected individuals often have motor deficits such as delayed motor development, hypotonia and ataxia. Approximately 30% of these patients present with cleft lip and palate, cardiac anomalies, orthopaedic and respiratory problems and also urinary structural abnormalities. The majority of patients with the classic Dandy-Walker syndrome are usually diagnosed within the first year of life. The symptoms tend to be related to hydrocephalus and increased intracranial pressure. Infants may present with weak neck muscles along with poor head control and spasticity. In older children, the clinical picture may involve delayed development, inability to walk and poor limb coordination. Seizures may occur in 20–30% of these patients. Delay in intellectual development is a common feature of the Dandy-Walker syndrome. The major clinical symptoms are neuromotor retardation (delay in normal neural and muscle tone development), mental retardation and hydrocephalus. Mental and intellectual functions are depressed in half of patients while other 50% may have normal cognitive functions.

The objectives of the surgical correction of macrostomia are accurate positioning of the oral commissure, reconstruction of a functional oral musculature and skin closure with minimally visible scar [5, 6]. It is generally agreed that the repair should be performed as early as possible to prevent feeding problems and aesthetic reasons.

Reconstruction of the oral commissure is an important part of surgical repair of macrostomia. Numerous surgical reconstructive techniques have been developed and used to normalize such defects. Examples are the straight-line closure, without supero-inferior rotation or positioning

of the scar [7]. We have used an inferiorly based square vermilion flap for the reconstruction of the angle of the mouth. Inferiorly based flap was chosen because of the abundance of vermilion found on the lower lip and also to create a smooth curve for better aesthetics.

Z plasty should be avoided for cheek closure because the scar will be visible in spite of meticulous suturing. Therefore, we used a lazy W-plasty for the cutaneous closure. In macrostomia, the orbicularis oris muscle is discontinuous and therefore it is important that the muscle ring be reconstructed. The sphincter-like function of the orbicularis oris muscle is very important both for feeding and for articulation. Reconstruction of the orbicularis oris muscle results in a good contour of the oral commissure and normal overlapping of upper lip.

Conclusion

The combination of a commissure plasty with a vermilion square flap from the lower lip and closure of the oblique facial cleft with a lazy W-plasty is an effective surgical procedure that yields consistent clinical result. Functional and well-contoured muscle reconstruction is the key to achieve good results in macrostomia repair.

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