Transverse Facial Cleft: An Uncommon Occurrence

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ABSTRACT

Background: Transverse facial cleft (TFC) is a rare facial cleft occurring at the angle of the mouth and runs in a horizontal direction posteriorly for variable distances. The increasing number of free cleft surgeries performed at various treatment centres across Nigeria sponsored by Non-Governmental Organizations suggests that practicing cleft surgeon will invariably encounter one or more rare variants of facial clefts such as transverse facial clefts. The objective of this paper is to review available literature on the aetiology, classification, presentation and surgical management and to document our experience with transverse facial cleft in our practice.
Methods: A literature review was conducted using Pubmed and Google scholar databases. It concentrated on manuscripts and overviews published in the last ten years. The pattern of presentation and surgical management of patients who presented to our practice over a period of 3 years were also discussed.
Results: Four patients aged 4months, 6months, 7 and 17years were managed in the period under review. Two of the patients presented with bilateral TFCs with the other two presenting as unilateral TFCs with no syndromic association. All four patients were managed surgically using a straight line repair technique.
Conclusion: The rarity of transverse facial cleft has made it difficult to design a specific treatment method however, despite the controversy over the preferred repair technique, good aesthetic and functional outcome was observed using the straight line repair technique. in 4 cases treated in our centre.

Keywords: Macrostomia, Transverse facial cleft, Tessier 7, straight line repair.

INTRODUCTION

Transverse facial cleft (TFC) also referred to as lateral facial cleft, congenital macrosomia, prosopoanoschisis, macrostoma is a cleft occurring at the angle of the mouth and runs in a horizontal direction posteriorly for variable distances depending on severity (Oghale & Chris-Ozoko 2013). It can be unilateral or bilateral and corresponds to Tessier 7 cleft on the Tessier classification. It may be a solitary finding or be combined with other clefts and/or syndromes.

The increasing number of free cleft surgeries performed at various treatment centres across Nigeria sponsored by Non-Governmental Organizations suggests that a practicing cleft surgeon will invariably encounter one or more rare variants of facial clefts such as the transverse facial clefts.

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The objective of this paper is to review available literature on the aetiology, classification, presentation and surgical management of transverse facial cleft and to report our experience in our practice.

MATERIALS AND METHODS

A literature review was conducted using Pubmed and Google scholar databases. It concentrated on manuscripts and overviews published in the last ten years. The key terms employed were "horizontal cleft", "Tessier 7 cleft", "transverse facial cleft", "lateral facial cleft", "macrostomia", "macrostomia repair", "surgical treatment transverse facial cleft", "oculo-auriculo-vertebral-spectrum", "Goldenhar syndrome", "Treacher-Collins syndrome" and "hemifacial microsomia". A review of the aetiology, classification, presentation and surgical management of transverse facial cleft was done. In addition. We reported our experience with TFCs, surgical technique used and follow up of four (4) patients.

AETIOPATHOGENESIS

Different theories have been put forward to explain the aetiology of transverse facial cleft (Veau, 1938; Gorlin et al.,1990; Anantanarayanan et al., 2007; Ahmed et al., 2010; Okpokowuruk & Amanari, 2013). The Dursy-His proposed that the face is formed from fusion of five facial processes – the frontonasal, paired maxillary and paired mandibular. Failure of epithelial fusion theory proposed that failure of fusion of the epithelium of these processes is believed to cause the formation of facial clefts (Gorlin etal., 1990). The mesenchymal penetration theory suggest that a deficiency in mesenchymal penetration of the embryonic facial region leads to the formation of facial cleft ((Veau, 1938).

Specifically, TFCs are believed to result from failure in the fusion of the mandibular and maxillary processes of the first branchial arch Anantanarayanan et al., 2007; Okpokowuruk & Amanari, 2013). Different aetiologies have been implicated. Genetic factors include mutations, variations and deletions involving various genes such as the cleft lip and palate transmembrane protein 1, GAD 1, IRF6 gene, MSX 1, PVRL1 (Leslie & Marazita, 2013).

Vascular cause has been attributed to an embryonic haematoma formation in the stapedius artery system. Damage or interruption of the stapedius artery results in inadequate arterial supply of the first branchial arch (Poswillo, 1973). Also implicated are environmental factors that includes infectious diseases, nutritional deficiencies, drugs and chemicals that induce embryonic dysmorphogenesis and uncontrolled apoptosis (Anantanarayanan et al., 2007; Oghale & Chris-Ozoko 2013). The aetiology of transverse facial cleft is however not clear.

CLASSIFICATION

Various classification systems have classified facial clefts using anatomical location, syndromic association, anatomical structures, and laterality. While anatomical classifications are based on bony landmarks of the craniofacial skeleton, example is the Tessier classification, association with other clinical entities have been used to classify them as syndromic or nonsyndromic. The use of laterality simply classifies macrostomia as unilateral or bilateral.

The Tessier classification designed by Paul Tessier in 1976 assigns a specific number to the site of each craniofacial malformation based on its relationship to the sagittal midline of the facial bone (Tessier, 1976). It relates soft tissue to skeletal landmarks and divides the face into an upper portion (cranial clefts) and a lower portion (facial clefts) based on the orbits. Fifteen locations for clefts were classified. Based on the Tessier classification, transverse facial cleft was assigned the Tessier 7.

Although, the clinical expression of a Tessier 7 cleft is highly variable, a severe Tessier 7 cleft begins as a macrostomia at the oral commissure and continues as a furrow across the cheek towards a microtic ear. The fifth and seventh cranial nerves and the muscles that they supply may be involved. The osseous component involves the zygomaticotemporal suture with hypoplasia of the zygoma and temporal bone. The zygomatic arch is disrupted and represented by proximal and distal stumps; varying degrees of mandibular deficiency, including complete absence of the ramus on the affected side.

In addition, transverse facial has also been classified as either unilateral or bilateral, syndromic, or non-syndromic, simple or complex. A recent study proposed a new classification of transverse facial cleft based on the direction of the anatomical appearance. T7.1 for superiorly rotated lateral clefts, T7.2 for middle positioned lateral clefts, T7.3 for inferiorly rotated lateral clefts and finally T7.4 for the agenetic type (Butow & Botha, 2010).

Mohan et al. (2013) classified tranverse facial cleft based on anatomy but also added therapeutic implications. They classified transverse facial cleft into type I or minor unilateral macrostomia, where the cleft terminates medial to the anterior border of the masseter with cleft length between 1-2cm and its repair requires vertical reorientation of the orbicularis oris muscle. Type II or major unilateral macrostomia was subclassified into IIa, cleft extends sagittally to the tonsillar pillars and IIb, cleft extends distal to the anterior border of the masseter and up to the tragus. This requires

masseter reconstruction in addition to orbicularis oris reorientation. Type III or bilateral minor macrostomia is similar to type I but bilateral and here the surgeon must define the commissural position without the aid of a normal contralateral side. Type IV or bilateral major macrostomia is subclassified into a, and b, similar to type II but bilateral.

EPIDEMIOLOGY

Transverse facial cleft is a very rare occurrence. Various studies have reported different incidence rates. Rances et al. (2015) reported an incidence of 1.43-4.85 per 100000. Oghale & Chris-Ozoko (2013) reported an incidence of 1:50000 to 175000 with an occurrence of 0.7-5.4 out of 1000 cases of cleft lip while Okpokowuruk & Amanari (2013) reported an incidence of 1:60000 to 300000. Unilateral transverse facial clefts are believed to be more common than bilateral and syndromic association is less common when compared to nonsyndromic cases (Oghale & Chris-Ozoko, 2013; Okpokowuruk & Amanari 2013).

CLINICAL PRESENTATION

The clinical presentation of transverse facial cleft depends greatly on whether it is unilateral or bilateral and/or association with other craniofacial deformities or syndromes. They usually present with macrostomia also called "fish mouth". Where it is associated with other craniofacial deformities, they usually present with ear anomalies like low set ears, preauricular tags, flattened nasal bridge, hypertelorism and high arched palate (Ahmed et al., 2010; Okpokowuruk & Amanari 2013).

Syndromes associated with macrostomia include Ablepharon-Macrostomia Syndrome, Barber-Say syndrome, hemifacialmicrosomia, branchial arch syndrome. ((Jackson et al., 1988; Akinmoladun et al., 2007; Okpokowuruk & Amanari 2013). Macrostomia can present as simple (partial) where the wide oral aperture with loss of commissural anatomy extends to the anterior border of the masseter or complex (complete) with full thickness defect of the face extending from the mouth to the tragus. In macrostomia, the defect affects the skin, orbicularis oris muscle and oral mucosa.

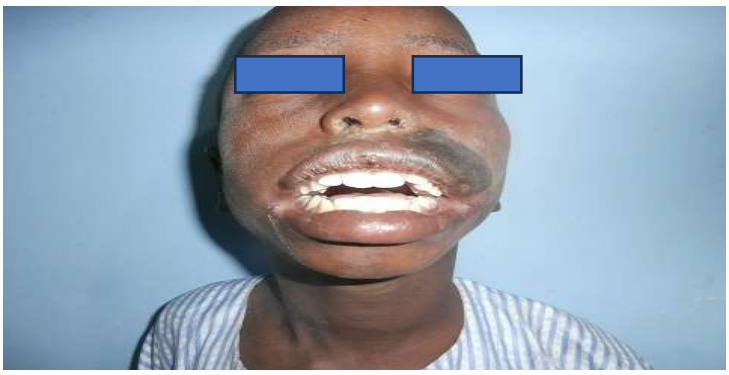

Figure 1: A 7year old boy with non-syndromic bilateral transverse facial cleft.

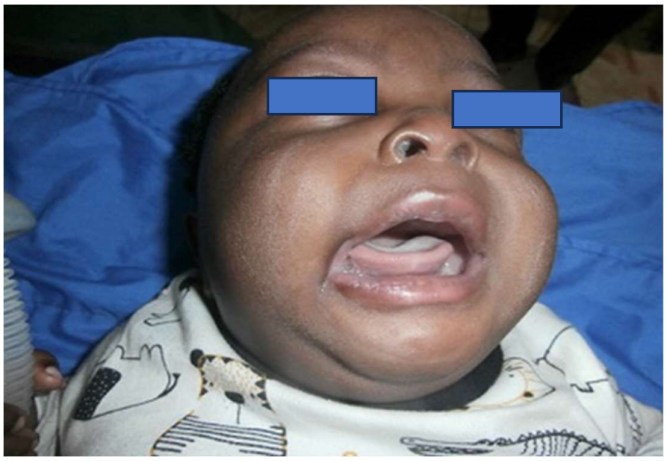

Figure 2: A 2-year-old boy with non-syndromic right unilateral transverse facial cleft

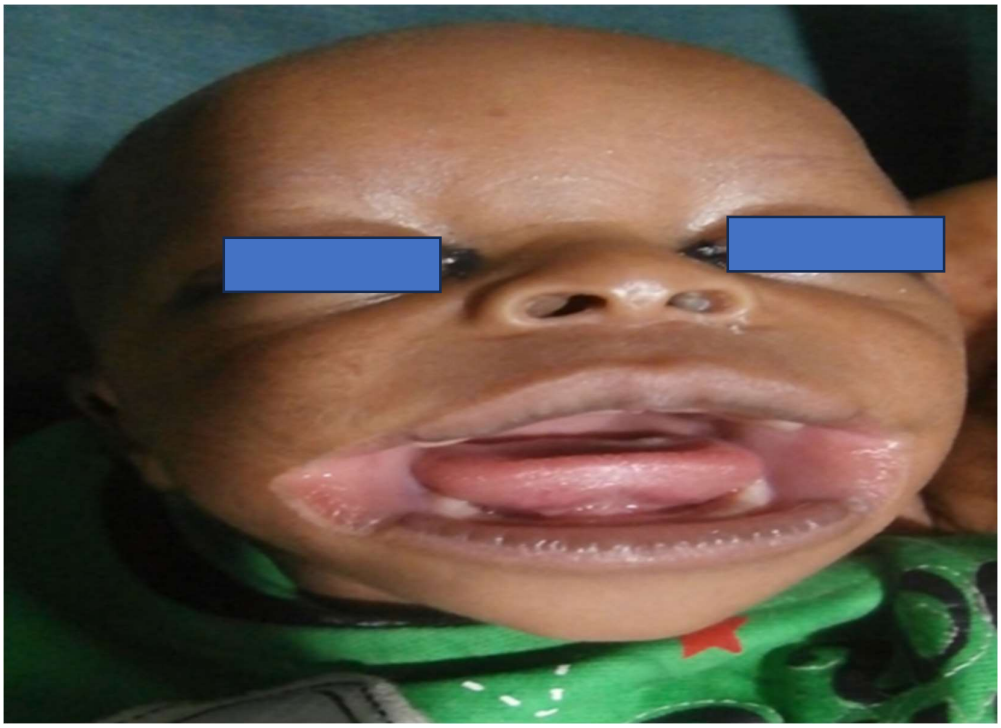

Figure 3: A 12 month old boy with non-syndromic bilateral transverse facial cleft.

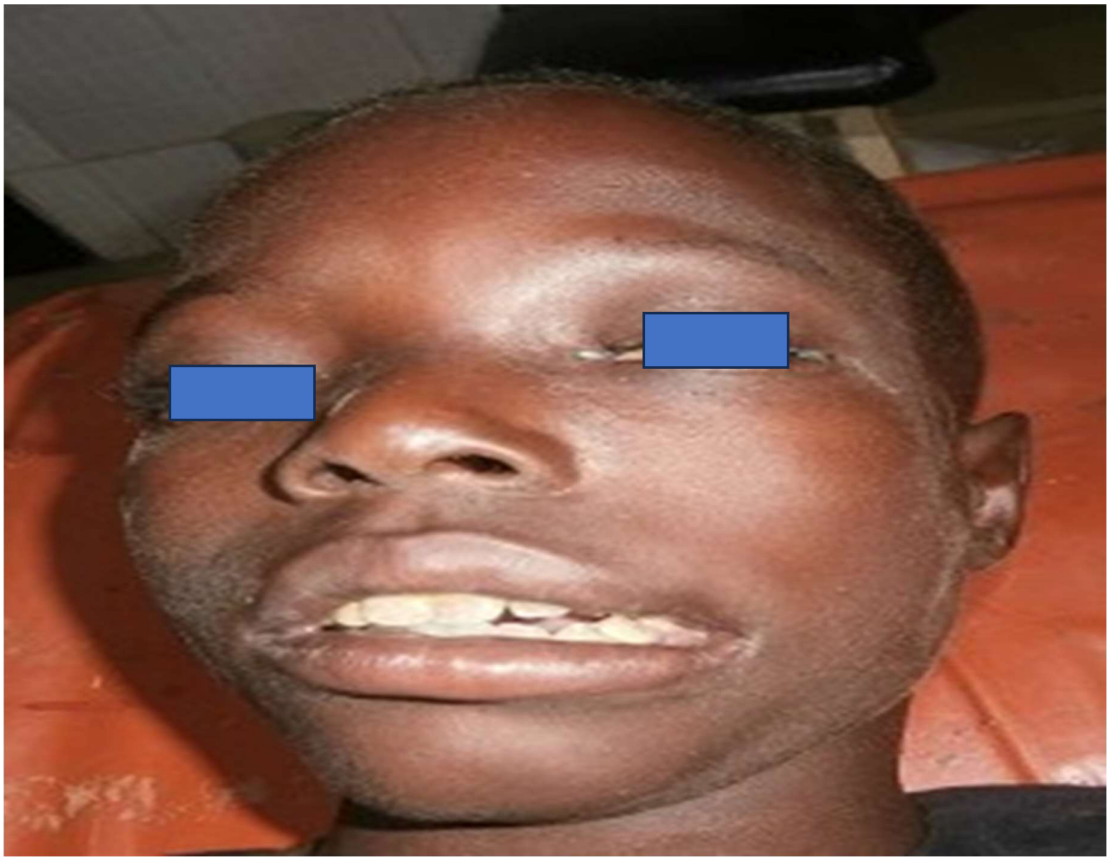

Figure 4: A 17 year old boy with left unilateral non-syndromic transverse facial cleft.

Figure 5: Postoperative photograph showing repair of unilateral (right)

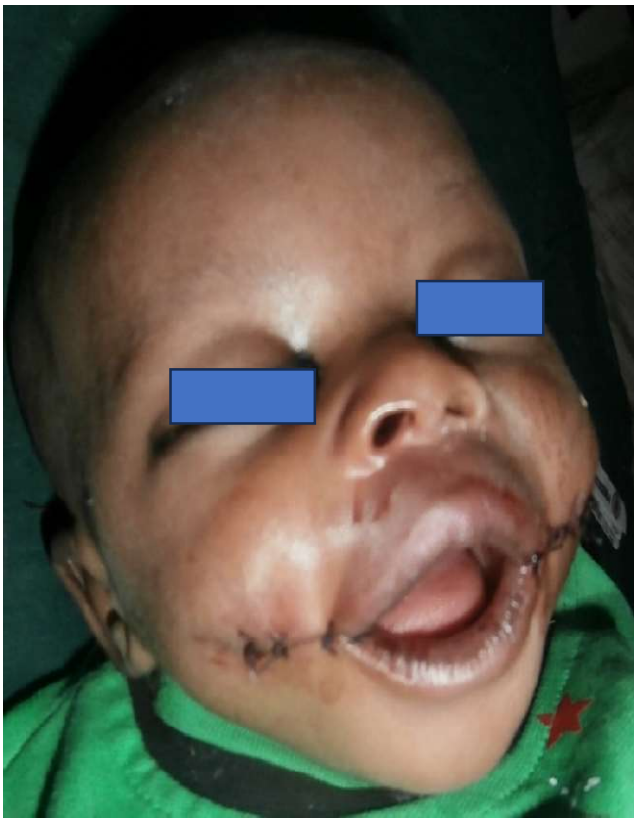

Figure 7: Photograph showing repair of bilateral transverse facial cleft

Figure 8: Postoperative photograph showing repair of left unilateral transverse facial cleft using the straight line repair technique.

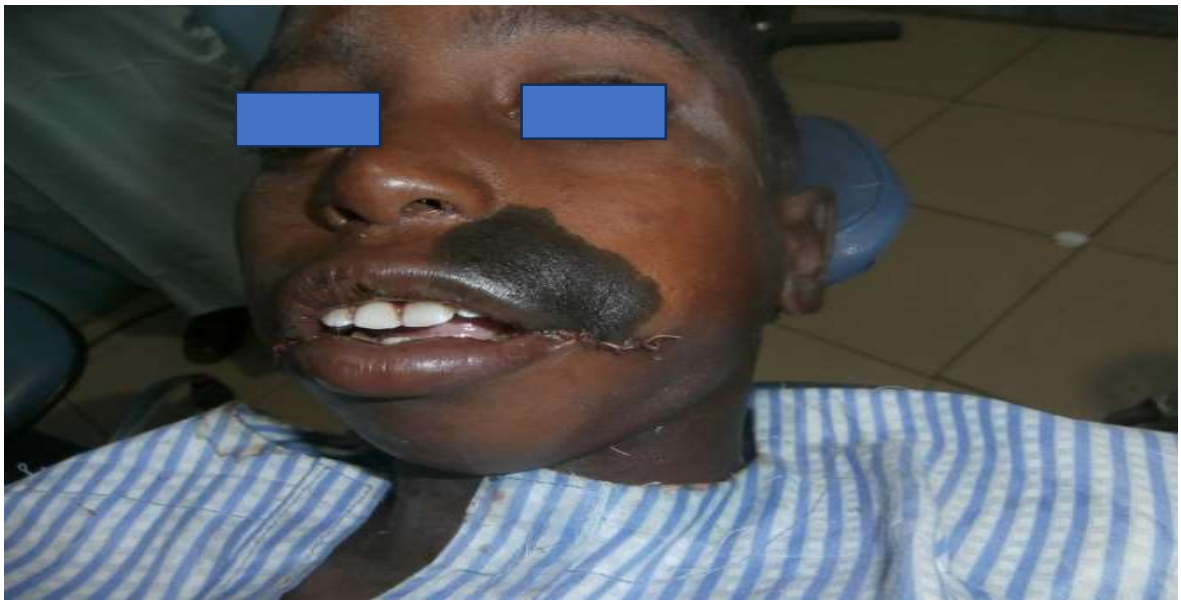

Figure 9a: Postoperative cleft repair in a 7year old showing the left side

Figure 9b: Postoperative cleft repair in a 7year old showing the right side

Table showing a list of some syndromes associated with transverse facial cleft

S/NO	SYNDROMES	COMPONENT
1.	ABLEPHARON-MACROSTOMIA SYNDROME	Absent or underdeveloped eyelids, macrostomia, low set ears with attached earlobes, syndactyly, bulging cheeks.
2.	BARBER-SAY SYNDROME	Hypertrichosis, fragile (atropic) skin, ectropion, macrostomia.
3.	HEMIFACIAL MICROSOMIA SYNDROME/ GOLDENHAR SYNDROME/ BRANCHIAL ARCH SYNDROME	Craniofacial anomalies in association with vertebral, cardiac, renal and central nervous system defects.
4.	BECKWITH-WIEDEMANN SYDROME	Macrosomia, macroglossia, omphalocele, embryonal tumours, ear creases and pits, renal abnormalities and adrenocortical cytomegaly.
5.	PERLMAN SYNDROME	Polyhydramnios with neonatal macrosomia, nephromegaly, renal dysplasia, nephron-blastomatosis, and predisposition to Wilmstumor.
6.	TREACHER COLLINS SYNDROME	Downward displacement of the lower eyelids, hypoplasia of the mandible and zygoma, dental anormalies, ear deformities and occasionally macrostomia

DISCUSSION

The treatment for transverse facial cleft is early surgical correction. The goals of repair include the symmetric placement of the neo-commissure, restoration of oral competence by repair of orbicularis oris muscle and closure of buccal mucosa to achieve a normal contour and prevent lateral migration of the commissure.[15]Each case of

macrosomia needs to be assessed with every surgical plan tailored to achieve the best results.

We have seen four (4) cases of transverse facial clefts in North-West Nigeria over an 8 year period. Total number of cleft patients seen and treated over this period is 1368 giving a prevalence of about 0.29%. As presented here, we saw two (2) unilateral and two (2) bilateral transverse facial clefts. Ages at presentation varied from childhood to teenage years. Apparently, most practicing cleft surgeons are not familiar with surgical repair of the transverse facial cleft lip, whether unilateral or bilateral. Repair of these clefts is similar to typical clefts in the sense that a three-layered (mucosa, muscle and skin) repair is necessary

For the unilateral transverse facial cleft, the horizontal plane of the commissure on the non-cleft side is a guide to locating the new commissure in the vertical axis (with the head positioned such that the alar-tragal line is parallel to the floor). In the bilateral transverse facial cleft, one of the surgical aims is to ensure that both commissures are in the same plane on the horizontal axis. It is also important to ensure that the upper lip is relaxed and provides full coverage of the upper teeth.

Muscular re-adaptation is another important aspect of surgical repair for transverse facial clefts. Typically, the muscle is some distance away from the edge of the cleft. The muscles need to be dissected out and reconnected (just as in typical cleft lip repair). Finally, attention should be placed on the mediolus (normal confluence of various muscle groups at the commissure of the mouth). The criss-cross pattern of muscles at this junction can be reproduced by a well-placed Z-plasty just before the commissure.

Many procedures have been explained in literature for surgical repair of macrostomia (Khorasani et al., 2019; Eguchi et al., 2002; Ahmed et al., 2010; Fadeyibi et al., 2010). All techniques recommend a layered repair starting with closure of mucosal layer, reconstruction of muscle layer before final skin closure. A straight-line closure is usually used to repair the inner mucosal layer as reported in most literature. The muscle layer is repaired usually by duplicating the orbicularis oris muscle with the upper branches overlapping the lower branches.

Although, there is controversy on the preferred technique for skin closure, the Z-plasty and the W-plasty are the most used techniques. A straight-line technique has also been reported, however, the Z-plasty is believed to be associated with less scar hypertrophy, contracture, and migration of commissure. Khorasani et al. (2019) reported excellent aesthetic and function results for 5 cases of transverse facial cleft which was repaired using the Butow& Botha technique. This technique is based on the placement and appearance of the transverse cleft. This technique essentially involves a four layered closure with a superiorly based vermillion flap and a modified cutaneous z-plasty rotated superiorly for skin closure. The essence of this is to achieve normal facial expressions during smiling.

CONCLUSION

The rarity of transverse facial cleft has made it difficult to design a specific treatment method however, despite the controversy over the preferred repair technique, good aesthetic and functional outcome has been reported for different surgical technique.

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Conflicts of interest

None declared

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