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REVIEW

# Diprosopus: a review of the aetiology and case report of early surgery in a 7-week-old infant with partial facial duplication

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## Abstract

Diprosopus is a rare craniofacial anomaly and is considered a subgroup of conjoined twinning. It encompasses a broad spectrum of duplications from single structure doubling to two completely formed heads on one neck. The aetiology of diprosopus remains a controversial topic, and many hypotheses exist. A multifactorial causal relationship is feasible because of the coexistence of other internal system anomalies of cardiac, gastrointestinal, neurological and respiratory origin. Ovid and PubMed databases were searched, using the key words: "diprosopus", "craniofacial duplication" and "duplicated mandible". The identified literature and key referenced articles therein were examined. The purpose was to consolidate the existing body of knowledge on the aetiology and management of diprosopus in order to improve our understanding of this rare condition. Our case report is relatively unique in that it represents a complete duplication of the mandible. Other tissues that were duplicated include the tongue, floor of mouth and the lower lip. It was hypothesised that associated growth abnormalities of the facial skeleton could undergo spontaneous correction if the duplicated mandible and associated tissues were excised early. On review, surgery is the only treatment that can offer functional, aesthetic and psychological improvement. Most patients who were offered surgery (in the reviewed literature) had a partial facial duplication. Surgery is usually deferred until the patient is older when more soft tissue is available to perform the reconstruction and when there would possibly be less growth restriction due to growth centre manipulation. This finding on the timing of surgery was in direct conflict with our hypothesis.
Level of Evidence: Level V, therapeutic study.

**Keywords** Diprosopus · Duplicated mandible · Aetiology · Management · Craniofacial duplication

## Introduction

Diprosopus is a form of conjoined twinning, resulting in a congenital craniofacial anomaly, in which any degree of duplication of the face occurs. The most severe end of the spectrum is an individual with one body and two faces [1, 2].

The word diprosopus comes from the Greek words di, meaning "two" and prosopon, meaning "face" [3, 4]. The global frequency of conjoined twinning is 1.2 per 100,000 births [5]. The prevalence of diprosopus is 1 in 15,000,000 births [3, 6], making it the rarest type of conjoined twinning (0.4% of conjoined twins) [3, 7]. In an epidemiological study by Martínez-Frías et al., raw data obtained from Castilla et al. was analysed and the incidence was calculated to be 0.06 per 100,000 births. [5] In diprosopus, the female-to-male sex ratio is considered to be 1.3:1 [8]. This number has been challenged by Martinez-Frias et al., who state that there actually may be more males than females affected. They recommend further studies due to the small sample size [5]. In conjoined twins, the incidence is higher in females. Two hypotheses have been suggested for this. One is that of abnormal X-chromosome inactivation contributing to the formation of conjoined twins [9]. The other is that male foetuses may be at higher risk for spontaneous abortion, as is seen in Trisomy 18 [9]. Stillbirths occur in 50% of all conjoined twins [10]. In one third of those born alive, severe abnormalities are present for which surgical intervention is not possible [10, 11]. Sixty-percent of those born alive do not survive long term [10, 12]. However, partial facial duplication can be compatible with life, and

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the surgical treatment is dependent on the nature of the deformity [3].

The diprosopus phenotype encompasses a wide spectrum of duplications [6, 7, 13–15], ranging from duplication of one craniofacial structure, i.e. *partial* craniofacial duplication [1, 4, 8, 16, 17] (involving only the face, cranial components or a combination thereof), to two complete faces on one neck [1, 4, 6, 8, 16, 18] or, in other words, a *complete* duplication of craniofacial structures [4, 17] (diprosopus monocephalus) [19]. The diprosopus spectrum can be associated with a variety of other anomalies, especially complete diprosopus [20]. According to Wu et al., in patients with partial duplication, the mandible and mouth are most commonly affected [21]. It is difficult to determine accurately the ratio between mandibular vs. maxillary duplication. It is important to mention that Shaikh et al. [22] has been misquoted in a number of articles as saying that maxillary or midface duplication occurs more commonly than mandibular duplication [4, 15, 23, 24].

The purpose of this article is to report a case of a 7-week-old infant with a duplicated mandible and associated oral anomalies that underwent a two-stage surgical correction in early infancy. The literature is also reviewed with an emphasis on the aetiology of diprosopus.

## Materials and methods

Ovid and PubMed databases were searched for articles reporting on human studies only. The type of article and the time period for the search were not specified. The key words used were: "diprosopus", "craniofacial duplication" and "duplicated mandible". The identified literature and key referenced articles therein were examined to prepare a collective review of the aetiology.

### Aetiology

The pathogenesis of diprosopus is controversial and is yet to be understood fully [1, 25]. Many hypotheses exist to explain the initiating event, but most researchers agree that the notochord, which induces neurulation, is at the heart of the problem [2, 14]. Each human embryo (and vertebrate) must have its own notochord [2]. As diprosopus is regarded as a form of conjoined twinning [15], the events that lead up to twinning are also considered as possible contributors to the condition [16]. Between days 13–25 of gestation, as the primitive streak and axial structures start to form, the normal development of the notochord is disturbed and diprosopus may ensue [20, 26]. The notochord is duplicated in these patients, and the two main postulations for this condition are either incomplete rostral splitting/fission of the notochord [4, 6, 13, 20, 21, 26–29] or fusion of two separate but adjacent notochords at susceptible locations [2, 26]. The theory of fusion is conceptually supported by the similarities demonstrated by the specific site of fusion in craniopagus and pygopagus twins [2]. Fusion commonly involves structures in the vicinity of two neuropores [2]. Also, Logrono et al., quoted by Kaufman, performed DNA typing on parasitic conjoined twins and found dizygosity instead of monozygosity [30].

Duplication of cells at the anterior termination of the notochordal process may initiate duplication of facial-oral elements [28] (Figs. 1 and 2).

Kaufmann does not regard fusion as a possible cause of conjoined twinning [19, 30, 31]. Machin suggests that diprosopus is the only condition where there is true rostral notochord bifurcation, compared to other conjoined twins where there are two independent

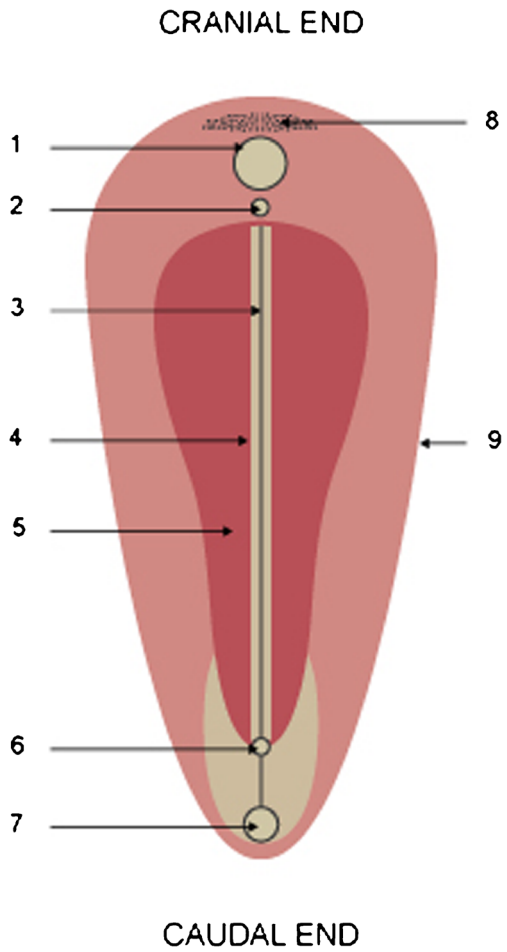


**Fig. 1** Normal longitudinal schematic image of a human embryo with a single notochord. 1. Oropharyngeal membrane, 2. Prechordal plate, 3. Neural groove, 4. Notochord, 5. Neural plate, 6. Primitive node, 7. Cloacal plate, 8. Cardiac anlage, 9. Cut edge of Amnion

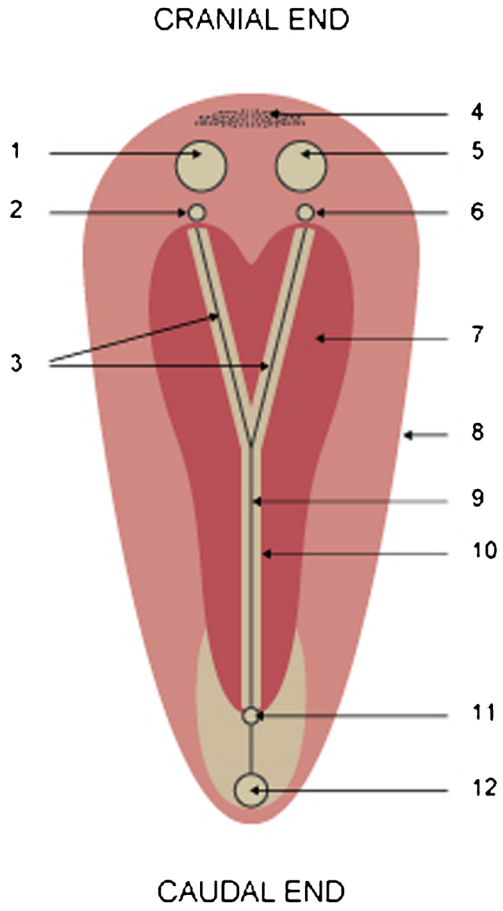


**Fig. 2** Longitudinal schematic image of a human embryo showing bifurcation of the cranial portion of the notochord. 1. Oropharyngeal membrane, 2. Prechordal plate, 3. Anterior duplication of the notochord, 4. Cardiac anlage, 5. Oropharyngeal membrane, 6.Prechordal plate, 7. Neural plate, 8. Cut edge of amnion, 9. Neural groove, 10. Notochord, 11. Primitive Node, 12. Cloacal plate

notochordal axes [32]. He also puts forward "interaction aplasia" as a possible explanation for why some tissues or organs fail to develop in these cases. This could be due to conflicting cell movement routes or aberrant foci of morphogens [32].

In partial diprosopus, only certain craniofacial elements or components of the first branchial arch are duplicated [4, 15, 17, 29, 33].

Possible regions of duplication are:

- The prosencephalon (when there is duplication of the eyes and nose),
- The olfactory placodes,
- The maxillary and/or mandibular growth centres around the stomatodaeum [31, 34].

Although pathogenetically unusual, bilateral components may occur in a unilaterally positioned duplicated structure around the stomatodaeum [29].

There does not seem to be a single cause, but a multifaceted series of events that leads to the recognisable phenotypical spectrum of diprosopus [28]. Potential causes are related to the foetal environment [35], the uterus [30], the condition of the ovum [36], abnormal placental circulation [8], teratogen exposure [37, 38], abnormal neurocristopathy [24, 39–42] and genetics [8, 13, 43]. Amniotic band syndrome (ABS), which has been associated with clefting and other anomalies (like anencephaly) [23, 44], has also been implicated as a cause [41]. However, documented cases of craniofacial duplication with ABS have not been reported [31].

Stockard experimented with the temperature and oxygen concentrations of trout embryos just prior to gastrulation and induced conjoined twins, including diprosopus-like embryos, in 1–2.3% [35]. The foetal environment is not the only factor that seems to play a role as there have been documented twin pregnancies where only one foetus had diprosopus [16, 34, 45].

Irregular or extended menstrual cycles in young or perimenopausal females may lead to late ovulation with over-ripening of the ovum, increasing the risk of duplication of the blastomere [36]. Maternal protein and riboflavin deficiencies may cause multiple ovulation or

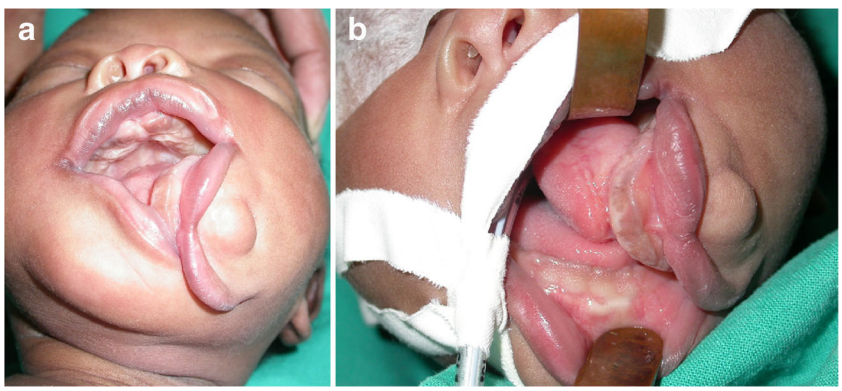


**Fig. 3** **a** and **b** A duplicated mandible is shown on the left-hand side of the oral cavity. The duplicated mandible is seen lying vertically and features its own floor of mouth, duplicated tongue and lower lip (on the right of the figure)

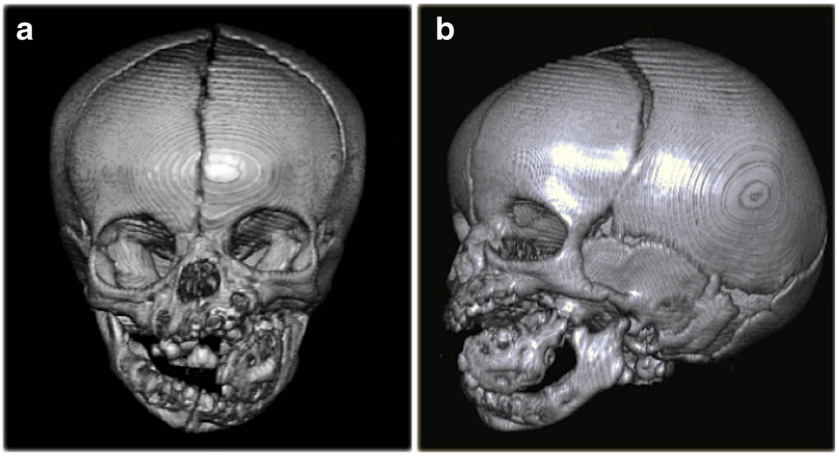


**Fig. 4** **a** and **b** CT scan and 3D facial bone reconstruction showing a left-sided deformation of the maxilla and primary mandible with an associated open bite, resulting from an unattached space-occupying duplicated mandible in the oral cavity. The left orbit is positioned higher than the right and the temporo-mandibular joint on the left appears to be normal. The duplicated mandible shows numerous non-erupted teeth

polyovular follicles or, otherwise, cause delayed implantation in the endometrium [36]. Conjoined twins may also occur after assisted reproduction [30, 46]. The condition of the uterus may be affected by the recent use of the oral contraceptive pill, possibly leading to delayed implantation, which is also considered as a contributor to diprosopus development in itself [30, 35]. It has been noted that there is an increase in the incidence of monozygotic twinning in patients where pregnancies followed within 6 months of cessation of the oral contraceptive pill, and a significant increase in congenital abnormalities when conception occurred within 3 months of stopping it [30].

It has been observed that there is a high frequency of monozygotic twins born to women with double monster offspring [47], creating the notion that there is a genetic component to conjoined twinning [8, 47]. There have also been familial examples of monozygotic twinning documented [30]. However, chromosomal analyses performed on diprosopus cases have been noted to be normal [21, 48].

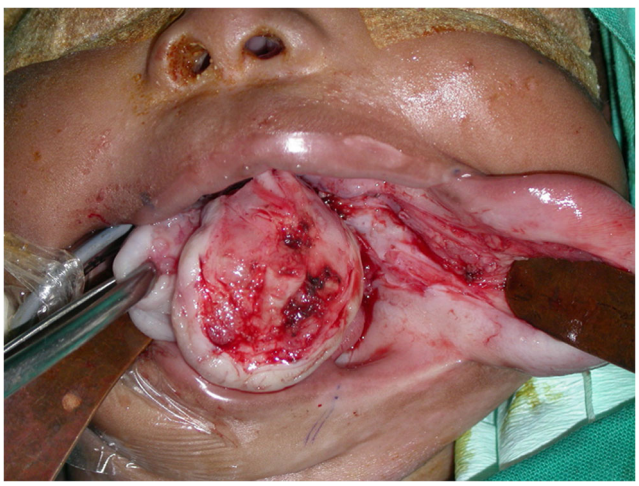

**Fig. 5** An intra-oral view of a near-total removal of the duplicated mandible and the tongue is shown, leaving behind some of the peripheral soft tissue floor

The inappropriate expression or deletion of homeotic genes (more precisely Dlx 5 and 6 homeobox genes) can lead to a large spectrum of anomalies across all germ cell layers [4, 16, 49, 50]. These genes are responsible for the spatial organisation of developing structures [16]. Mutant disorganisation has been studied in the murine model and is known to be transferred in a semi-dominant manner [51, 52]. Winter and Donnai suggest that there is a corresponding human disorganisation syndrome [52]. This syndrome has been linked to other cases of conjoined twinning and duplication of structures [26, 51]. Diprosopus may be a part of this spectrum.

The most common teratogen is ethanol [30]. In a hamster study, various other teratogens (vitamin A, dimethylsulfoxide, and urethan) induced twinning. All these twins also had some degree of failure of anterior neural tube closure [37]. Other teratogens that are implicated in twinning or neural tube anomalies are thalidomide, vincristine sulphate in mice, general and local anaesthetics, colchicine, griseofulvin and phenylzine dihydrogen sulphate [30, 37, 53]. A case of dicephalus has been associated with maternal exposure to Chernobyl in 1986 [12]. Other factors include viral and parasitic infections [54].

Neural crest cells are a fleeting population of multipotent precursor cells which then migrate and induce formation of diverse cell lineages [44]. Abnormal neurocristopathy may be caused by impaired cell migration or unsuccessful induction of structure-formation [39–41, 55], particularly craniofacial cartilage and bone formation [56]. Most defects seem to arise from failure of fusion of embryonic processes or failure of neural tube closure [5, 40].

Teratoma formation (sequestration of totipotent stem cells during early development with various tissue or

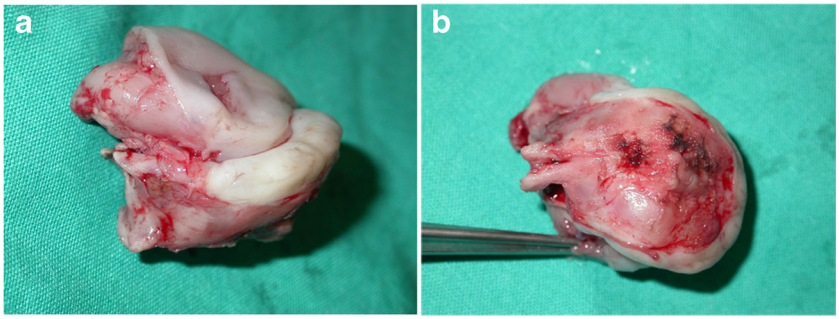


**Fig 6** **a** and **b** The superior surface of the excised duplicated mandible, floor of the mouth and tongue is shown (left). A posterior view of the duplicated mandible, the rami with its condyles abutting each other, i.e. posteriorly, the arch of the mandible is not open (right)

organ components) should be distinguished from partial duplication [17, 33].

## Case report

A twelve-day-old female neonate, born with duplicated congenital orofacial anatomy, presented to our quaternary centre, Inkosi Albert Luthuli Central Hospital (IALCH), in Kwazulu-Natal. Respiratory and feeding difficulties were not present. The antenatal history was unremarkable and no other congenital abnormalities were clinically obvious. To plan the surgical intervention, a computerised tomography (CT) scan with three-dimensional (3D) reconstruction of the head was performed. This was followed by an examination-under-anaesthetic to assess the abnormal duplicated oral structures. The duplicated structures included the mandible, tongue and floor of mouth, external soft tissues of the mentum and lower lip on the left. A cleft of the soft palate was also present (Fig. 3).

At the age of 7 weeks, surgical removal of the duplicated structures was accomplished (Fig. 4). The hypothesis that further deformation of the normal skeletal structures could be prevented and that growth correction of the maxilla, mandible and orbital position may occur with growth of the face was applied. Using an intra-oral approach, the duplicated mandible and tongue were excised from the vertically orientated duplicated floor of mouth. Soft tissues peripheral to the duplicated mandibular arch were retained. After the excision, the soft-palate cleft was better visualised and it was noted to be unusual as it was not central and the defect was on the left-hand side of the soft palate. A standard repair, therefore, was not possible (Fig. 5).

Instead, the residual soft-palate edge medial to the defect was pared (Fig. 6). A left palatopharyngeus musculo-mucosal flap, lateral to the defect, and mucosal

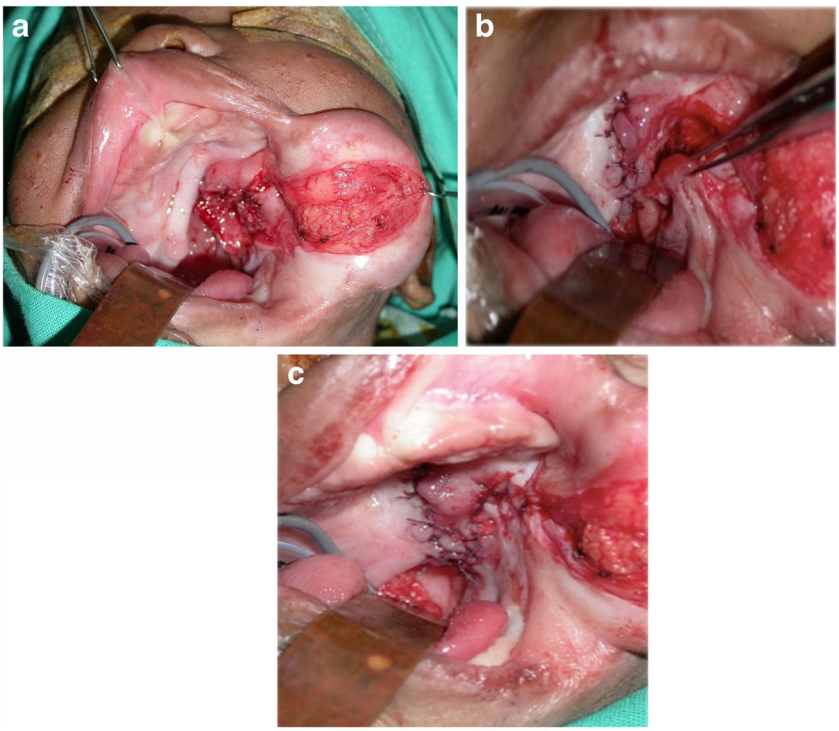


**Fig. 7** **a**–**c** The palatal defect is shown on the left-hand side of the soft palate after the medial edge is pared (**a**). A superiorly based palatopharyngeus flap is visualised, sutured to the medial side of the defect. The residual tissue of the duplicated floor is seen mobilised to close the lateral side of the defect (**b**). The soft palate defect is shown, sutured closed with the use of both flaps (**c**)

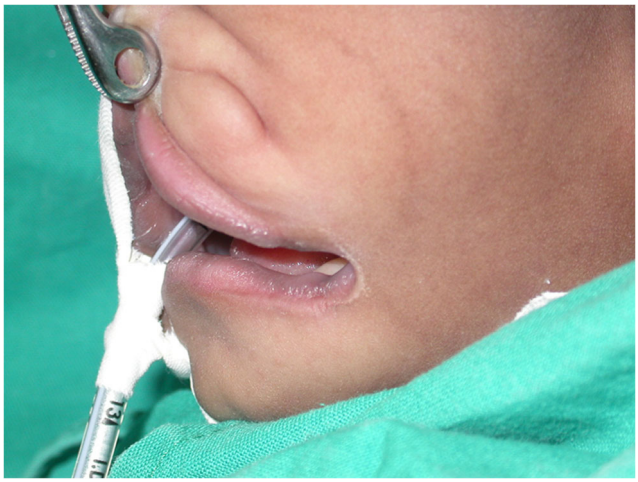

**Fig. 8** The junction between the duplicated lower lip (above) and the normal lower lip (below), simulating macrostomia of a Tessier No. 7 cleft

tissue from the residual duplicated floor of mouth was then rearranged to close the oral side of the defect in a dual layer. Flaps were laid parallel to each other (Fig. 7).

Following intra-oral defect closure, the external perioral tissues were addressed. To preserve a normal looking left commissure, the commissure between the normal upper lip and the duplicated lower lip was retained. The duplicated lower lip, 1 cm medial to this commissure with its associated soft tissues of the mentum, was removed. The duplicated lower lip, connected to the normal lower lip, resembled a Tessier's No. 7 craniofacial cleft, similar to macrostomia. The normal lower lip, inferior to the No. 7 cleft, was detached from the cleft, shortened, released and advanced to the cut edge of the duplicated lip near the left commissure and repaired. Excision of all soft tissues was performed as planned, closing the low-lying lateral oral cleft. Correction of lip aesthetics and oral competence was not our aim at this stage. Lip revision was to be considered once the hypothesised maxillary and mandibular deformity correction occurred in response to the removal of the duplicated structures (Fig. 8).

A minor complication of dehiscence of the palate repair was encountered post-operatively. This was allowed to heal spontaneously. The excised duplicated structures were analysed histologically. There were no tissue abnormalities noted. The duplicated mandible had representation of stratified squamous epithelium, odontogenic epithelium, bone, bone marrow elements and developing teeth (in keeping with a duplicated mandible).

Over the ensuing year, the mandible and maxillary deformities and their corresponding occlusion improved (Fig. 9).

At 17 months of age, the lip was revised with a wedge excision of the horizontally elongated lower lip, advancing it to the previously preserved left commissure. Lip competence and lip aesthetics were achieved. The patient was lost to follow-up at 1 month after the second-stage surgery (Fig. 10).

## Discussion

Diprosopus cases are challenging to manage and demand extensive interdisciplinary co-operation [19, 57]. The management approach is largely dependent on the nature of the duplication with only some partial duplications rendering themselves to appropriate surgical correction.

It is the ideal that this condition be diagnosed early on a detailed antenatal ultrasound. On ultrasound, specific findings can be highly suggestive of diprosopus [13, 58]. These include increased nuchal translucency, lymphangiectasia, neural tube defects, ventral wall defects, renal agenesis, a widened vertebral column, a

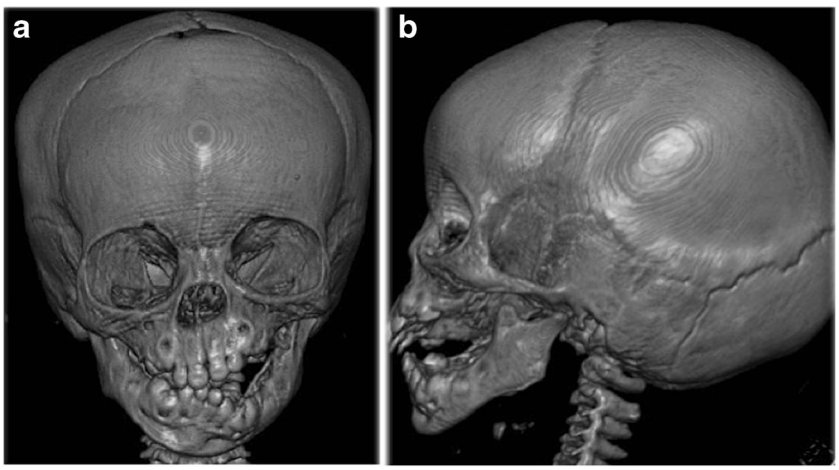


**Fig. 9** **a** and **b** The follow-up CT scan at 17 months of age shows compensatory growth of the affected maxilla and mandible, partially correcting the original deformity and dental occlusion present at birth. Orbital dystopia also has improved

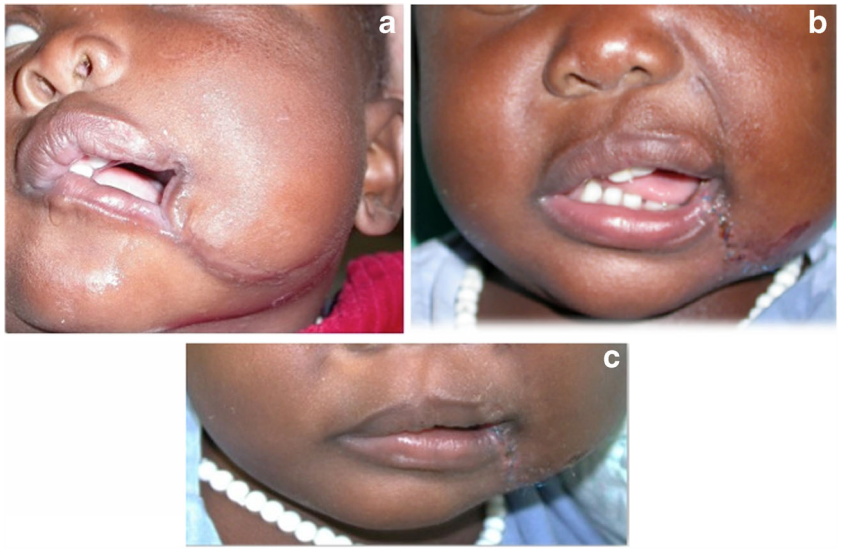


**Fig. 10** **a**–**c** Repair of the lip is shown at 6 months following the first stage operation (**a**). Repair of the lip is shown at the 1-week follow-up after the second stage operation at 17 months of age. An improvement in the open bite is also noted (**b**). The lips are shown apposed, demonstrating lip competence (**c**)

heart-shaped cranium or bifid cranial vault and duplication of the face or facial components [21, 46].

By diagnosing the condition earlier, a more thorough management strategy can be constructed, which would include delivery options, counselling, involvement of other disciplines and the decision to continue with the pregnancy if the parents so desire [3, 10, 58]. The parents should be counselled compassionately that there is a poorer prognosis with complete facial duplication [20] and that separation is impossible [11]. In an article by Thornton, the parents of a diprosopus baby opted to continue with the pregnancy and felt that with the support of the treatment team and comprehensive counselling, they were prepared to deal with the death of their baby very soon after delivery. It was much more peaceful than expected, and they were able to cope with their loss [57].

Isolated duplication, however, is not necessarily incompatible with life and some can often be managed surgically [30]. If the parents decide to continue with the pregnancy, they must be made aware that a caesarean section will most likely be required due to cephalopelvic disproportion [13]. Once the baby is born, a detailed management plan should be communicated and deliberated with the parents [56]. There are growth, functional and psychological considerations to weigh up when timing surgery [15, 18]. Surgery is the only treatment that can offer functional and aesthetic correction and will always be an improvement on the original, especially in severe deformities [15, 22].

Surgery has had varying success, and the parents should be made aware that there will be visible scarring [20, 21, 29, 34, 53, 56]. Obwegeser was very critical of his scars post-operatively despite multiple revisions [22, 56]. However, Price is of the opinion that good results can be expected and that surgery is mostly straightforward [59]. Surgery is usually deferred until the patient is older and more soft tissue is available to perform the reconstruction. Obwegeser waited until the patient was 10 years old but still found that the midface had growth retardation [56].

In our case, we opted to operate as early as 7 weeks of age on the supposition that removing the intra-oral duplicated mandible and tongue would eradicate the forces of deformation, produced by their on-going growth, on the rest of the non-duplicated craniofacial skeleton. Although there were no feeding and no airway problems at presentation, they may have developed as the child grew were these duplicated structures not removed. Also, asymmetry of the face would have worsened, sacrificing the psychological well-being of the growing child and parents. The interval between the first stage and the second stage operations clinically and radiographically demonstrated a partial correction in the bony deformity of the facial skeleton. It was unfortunate that the patient did not return for further follow-up and that all means of contact failed. A longitudinal follow-up would have been invaluable.

Definitive surgical treatment of the perioral tissues was deferred to a later stage. As the skeletal deformity began to correct over time, it laid the foundation for an improved soft tissue lip positioning and scar contracture correction of the repaired Tessier No. 7 cleft. Maisels noted that when macrostomia was repaired, especially very early, there was a tendency for the commissure to drag as the child grew [15, 29]. We performed the surgical revision at the age of 17 months, but there could have been value in an even further delay, anticipating further correction of the jaws.

The soft palate cleft was atypical, presenting with a left-sided defect. The standard techniques of repair were not feasible. The cleft palate repair is generally deferred to 9 months of age or older. We purposely repaired the cleft of the soft palate during the excision surgery to enable the use of some of the redundant tissues of the duplication. If this opportunity had been missed, then a higher degree of surgical complexity, using local pedicled flaps or even free flaps, would have been required [60]. Duplicated mucosa excision and salivary apparatus removal was not performed, despite being recommended in the literature as there is a higher chance of developing retention cysts [14, 29, 33]. In our case, no cysts had developed up to the final review but, owing to the loss to follow-up, the risk could not be assessed thoroughly. Owing to this fact, long-term speech evaluation was also not possible. There was a paucity of information with regard to this in the literature. Eleven articles mention speech or speech outcomes in craniofacial duplication but they are not specific; the patients were not objectively evaluated [42, 61, 62] or no surgery was performed [63, 64]. The surgical cases highlight the fact that intensive speech therapy is necessary postoperatively to improve outcomes [42, 61]. The longest follow-up was noted to be by Stewart et al., which was 15 years [62]. The authors only state that their patient had good speech development. In the literature, it has been shown that better speech outcomes can be expected if the cleft palate is repaired prior to speech development [65], so we can surmise that there would have been an improvement in speech in our patient.

Long-term follow-up is important to address any residual deformities not corrected by on-going growth of the craniofacial skeleton. Potentially, orthodontic therapy, orthognathic and further lip revision surgeries are backup strategies to achieve an ultimately good result.

## Conclusion

Diprosopus is very rare. Its pathogenesis is multifactorial but abnormalities in the formation of the notochord may be central to its development. Mainly partial facial duplications are amenable to surgical correction. The type of surgery is dependent on the nature of the duplication. Early excision of the duplicated structures potentially interrupts deformational growth of the normal craniofacial skeleton and may even provide, to some degree, auto-correction. The surgical approach should include use of spare parts as well as strategic planning in the use of available soft tissues for the best possible functional and aesthetic outcome.

## Compliance with ethical standards

**Funding information** No financial disclosures.

**Conflict of interest** Sieglinde Erica Rabe, Mahendra Daya and Anil Madaree declare that they have no conflict of interest.

**Ethical approval** Ethical approval was obtained from the Biomedical Research Ethics Committee (BREC) at the University of Kwazulu-Natal, Durban, South Africa.

**Informed consent** There was no signed informed consent from the mother of the patient. The patient was from rural Kwazulu-Natal. The Social Worker of Stanger area was contacted and they were unable to locate the patient nor her mother. The ethics committee (BREC) was informed about this, and permission was granted by them to use the information and photos of the patient for my masters in medicine (MMed) degree (which also includes publishing the article). The letter is included in the supplementary forms.



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