Case Report

# A "three-eyed" infant: A case of partial facial duplication (diprosopus monocephalus triophthalmos)

Rabi Sani Yahaya, Mohammed Abba Suwaid[1], Sadiq Hassan, Yusuf Lawal[1], Ismail Hassan[2], Baffa Adamu Gwaram[3], Hadi Bala Yahaya[4]

Departments of Ophthalmology, [1]Radiology, [2]Surgery and [3]Medicine, College of Health Sciences, Bayero University, Aminu Kano Teaching Hospital, [4]Department of Ophthalmology, Murtala Mohammed Specialist Hospital, Kano, Nigeria

## Abstract

Craniofacial duplication known as diprosopus is a rare congenital disorder whereby parts or all of the face are duplicated on the head. This is a case of a 1-year-old boy referred to our hospital with an extra eye (third eye) on the left side of the head and an abnormally shaped head, which were noticed since birth. Pregnancy and delivery were uneventful. Apart from routine antenatal medication, there was no history suggestive of ingestion of traditional or other medication during pregnancy. No history of smoking during pregnancy and no history of exposure to radiation. The child was born in a rural area to a "nonconsanguineous marriage." Examination revealed posterior plagiocephaly, a depressed anterior fontanelle, and a bulging posterior fontanelle. There was an extra eye (third eye) in the left temporal region. Radiologic findings showed the presence of two normally situated bony orbits and two extra orbits in the left temporal region. Well-formed globes were seen in the normal orbits while only one of the extra orbits contained a globe. The importance of prenatal diagnosis is emphasized.

**Keywords:** Diprosopus, facial duplication, prenatal diagnosis, three eyes

**Address for correspondence:** Dr. Mohammed Abba Suwaid, Department of Radiology, Bayero University/Aminu Kano Teaching Hospital, Kano, Nigeria.
E-mail: drsuwaid@yahoo.com

## INTRODUCTION

Craniofacial duplication also known as diprosopus is an extremely rare congenital disorder whereby parts (accessories) or all of the face are duplicated on the head.[1,2]

It may be symmetrical or asymmetrical. Those cases that have been reported in the literature constitute a spectrum from simple nasal duplication to complete separation of two faces. There have been some reports of eye, nose, mandible, or maxilla duplication separately.[2]

Although classically considered conjoined twinning (which it resembles), this anomaly is not normally due to the fusion or incomplete separation of two embryos. The cause is unknown. It is, however, postulated to be the result of abnormal activity by the protein sonic hedgehog (SHH).[3]

Evidence has shown that the prevalence of diprosopus is 2/1,000,000 births. The facial structures more frequently duplicated are the nose and eyes. Another reported prevalence rate is 0.002/10,000 birth.[4] There is a predominance of females over males (2:1).[4] Frequently associated anomalies include anencephaly, duplication of cerebral hemispheres, craniorachischisis, oral clefts, spinal abnormalities, congenital heart defects, diaphragmatic hernia, and thoracic and/or abdominal visceral laterality anomalies.[5]

| Access this article online | |
|---|---|
| Quick Response Code: | Website: www.wajradiology.org |
| | DOI: 10.4103/wajr.wajr_33_17 |



**How to cite this article:** Yahaya RS, Suwaid MA, Hassan S, Lawal Y, Hassan I, Gwaram BA, *et al*. A "three-eyed" infant: A case of partial facial duplication (diprosopus monocephalus triophthalmos). West Afr J Radiol 2018;25:79-83.

Many human infants with diprosopus are stillborn. Few cases with this type of condition, however, have survived for longer periods.[5] Myths are commonly attached to such infants who are regarded as monsters and are often abandoned by their parents to avoid stigmatization. Hence, they rarely present to the hospital. A third eye with almost all its components is exceedingly rare,[2] with only a few cases reported in medical literature or published in newspapers.

We hereby present a case of a three-eyed male infant with a hidden fourth socket (diprosopus monocephalus triophthalmos) which is an extremely rare malformation with unknown etiology and reviewed the literature.

## CASE REPORT

A 1-year-old boy was referred to our hospital on account of having an extra eye (third eye) in the left temporal region and an abnormally shaped head, which were noticed since birth. The child was first taken to a peripheral hospital at the age of 9 days, but he was lost to follow up. Pregnancy was uneventful and the mother attended antenatal clinics. Apart from routine antenatal drugs, the mother did not ingest any type of medication (including traditional herbal medications) during pregnancy. No radiological imaging was done for the mother during pregnancy and there was no history suggestive of exposure to radiation. There was no history of illness or smoking. The pregnancy was taken to term; labor was uneventful and lasted 2 h. Delivery was at home, spontaneous, vaginal and supervised by a traditional birth attendant. The child cried immediately after birth. The mother noticed the head to be asymmetrical, with an abnormally placed eye (third eye) on the left side of the head. No history of vomiting and no seizures. There was a delay in achieving developmental milestones (he achieved neck control at 6 months and is still not able to walk or crawl at 1 year). He is not fully immunized for age. He is the fourth child of both parents in a monogamous setting from a rural area in Northern Nigeria. The parents are not related. Other siblings are alive and healthy.

Physical examination revealed a calm child, not pale, anicteric, afebrile, and acyanosed. The frontal hairline continued to the left temporal region, devoid of hair. There was posterior plagiocephaly with a depressed anterior fontanelle (1 cm × 2 cm) and a bulging and patent posterior fontanelle after 1 year (5 cm × 6 cm). There were no distended scalp veins. There was an extra eye (third eye) in the left temporal region [Figure 1]. Visual acuity for this could not be determined due to corneal haze and the inability to visualize other parts of the eye. Eyelids and lashes were vertically oriented and blinked spontaneously with the other eyes. The conjunctiva was injected and the cornea was hazy. As a result, other aspects of the globe could not be visualized. The right and left eyes were in the normal anatomical positions. The conjunctiva, cornea, and anterior chamber for each of the two eyes were normal. The lens for each eye was transparent with a good red reflex in each eye. Patient was uncooperative for a fundus examination. There was right facial nerve palsy. The cardiorespiratory systems and abdomen were essentially normal. Musculoskeletal and neurological examinations are normal.

The scanogram of computed tomography image showed mild craniofacial asymmetry, fullness of the left zygomaticotemporal region, and two roundish areas of lucency with sclerotic borders in the left para-orbital and adjoining zygomaticotemporal bony region. The native orbits, nasal cavities, and jawbones appeared unremarkable [Figure 2].

The axial contrast-enhanced images confirmed the previously noted left zygomaticotemporal lucent areas on the scanogram to be focal skull vaults fossa-like bony depressions with sclerotic margins [Figure 3]. One of the bony fossa-like depressions is covered by scalp tissue with no breach seen, whereas the other contained a fairly oval faintly marginal enhancing hypodense mass of soft-tissue density with an eccentric posterolateral located focus of hyperdense calcification [Figure 4a]. No significant vascular structural enhancement noted. The left parietal and occipital lobes appeared hypoplastic with a cerebrospinal fluid-filled cystic mass lesion noted posteriorly and a cranial defect. There is associated widening of the prepontine cistern presumably an arachnoid cyst [Figure 4b].

The remaining cerebral and cerebellar hemispheres including the ventricular systems of the brain were normal.

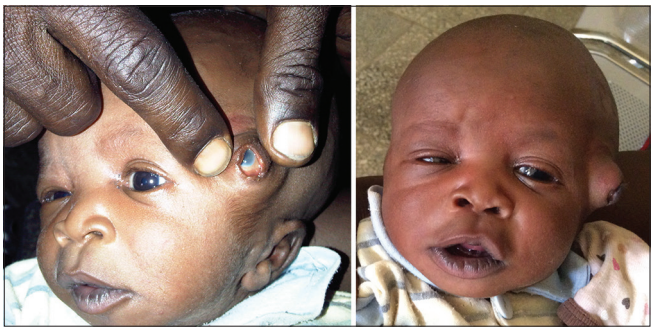

**Figure 1:** Clinical picture of the child showing the two normal located native eyes and an additional visible eye in the left temporal region

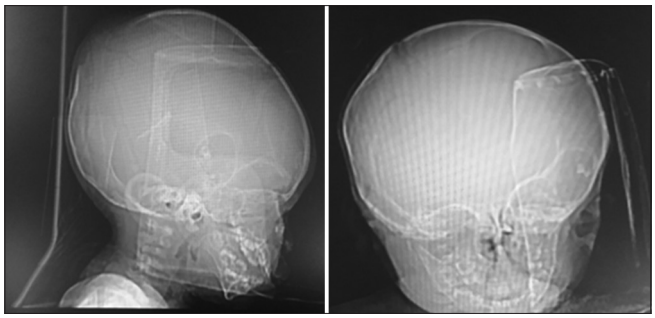

**Figure 2:** Scanogram anteroposterior and lateral showing two orbital cavity depressions in the left temporal aspect of the calvarium (arrows)

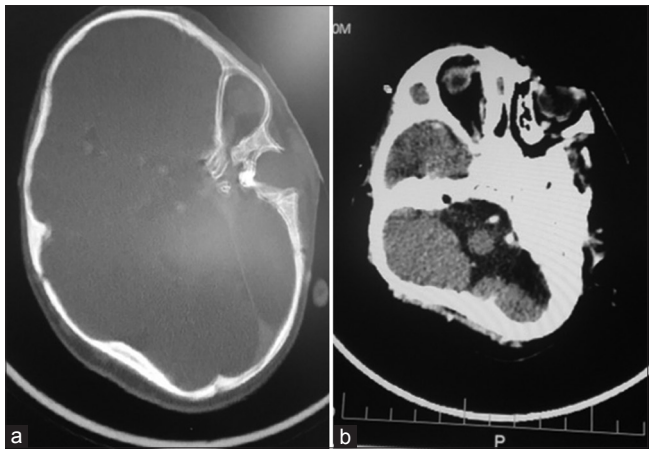


**Figure 3:** Axial computed tomography scan of the head showing two bony orbital cavities (vertical and horizontal double edged arrows) in the left temporal region (a) bone window (b) brain window

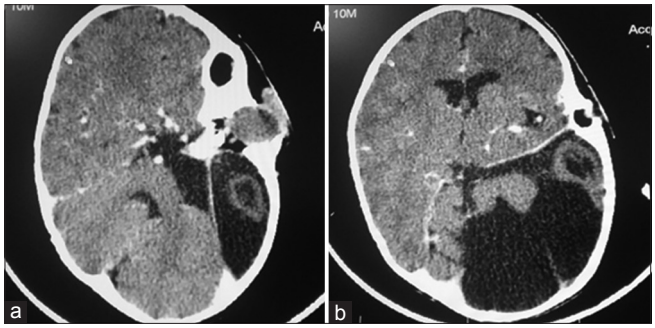


**Figure 4:** Axial computed tomography scan brain window showing (a) duplicated globe within the left temporal bony orbit (arrow), (b) a large arachnoid cyst in the posterior fossa (crossed arrows)

An magnetic resonance imaging could not be done as the patient did not return to the clinic.

## DISCUSSION

Conjoined twins have been reported in the literature since 1864; however, their existence has been known for centuries. The earliest known report on diprosopus has been credited to Ambroise Paré (of monsters and prodigies) of the 16th century.[6] Since 1884, there have been only 35 reports on diprosopus in the world medical literature.[7] To the best of our knowledge, there is no published case of postnatal diprosopus in Nigeria.

Singh *et al*. in Sokoto, North West Nigeria, reported only one case of diprosopus out of 10,163 deliveries.[8]

Diprosopus or craniofacial duplication is the rarest form of conjoined twinning, with an incidence of approximately 0.4% of all types of conjoined twins.[9] It is a Greek word meaning "two-faced person." The cause is unknown but it is postulated that this anomaly may represent arrest in uniovular twin formation. Protein SHH and its corresponding gene play an important role in signaling craniofacial patterning during embryogenesis. This protein governs the width of facial features.[3] The greater the widening, the more structures are duplicated, often in a mirror image form.

During embryonic development, SHH directs embryonic cells to organize in specific areas that later become specialized neural tissues, thus controlling the size and shape of brain structures. However, we could not measure SHH protein in the index case due to lack of requisite equipment and expertise.

The reported prevalence of 0.008/10,000 and 1% of all the monochorionic twins. They are classified mainly into three groups, terata catydidymus (Diprosopus, dicephalus, ischiopagus [6%–20%] and pyopagus [10%–20%]), terata anadidyma (dipygus, syncephalus, and crainopagus [6%–12%]), and terata anacatadidyma (thoracopagus [30%–40%], omphalopagus [25%–30%], and rachipagus).[4]

Other embryologic theoretical explanations that have been considered are either a "fusion" of two parallel notochords in close proximity occurs or "fission" of a single notochord occurs during the first few weeks after conception or duplication of neural crest cell derivatives and mutations of the *Dix* homeobox gene.[10] All these are, however, unproven theories as there have been no previous reports of genetic mutations associated with diprosopus. We could not carry out a genetic testing on the parents in this case due to the absence of requisite diagnostic tools. However, a lack of genetic association for diprosopus continues to support an embryologic theory of abnormal twinning.[9]

When a child with cranial vault deformity and swelling presents with delayed neuromuscular/developmental milestones, an encephalocele or arachnoid cyst is suspected but an accessory eye is a rare and unlikely consideration. Ingestion of certain medications and febrile illnesses during

pregnancy might increase the chances of craniofacial malformations.[5] Three eyes/accessory eyes are generally a rare form of craniofacial teratology characterized by the presence of additional orbital cavity/globe despite the presence of the normally located two native orbits and globes. This particular case presented with an arachnoid cyst. Apart from routine antenatal drugs, there is no history of ingestion of any kind of medication during the pregnancy. However, some of the food eaten may contain certain chemicals that may be implicated in this case. The subject has two additional orbits, with one orbit (third eye) having a well-formed globe, whereas the other (fourth orbit) being empty and completely covered by scalp tissue. Externally, the eye has eyelids and lashes that are well formed and are vertically oriented. Examination showed conjunctival injection, a hazy cornea while other aspects of the globe could not be visualized due to the corneal haze.

Mason in a review reported a case of a female infant with facial duplication that was born to a 15-year-old black primigravida. The case had a large nasofrontal encephalocele with four eyes, two noses, two maxillae, and one mandible. The two empty eye sockets were located in the interfacial region, immediately above the mouth and overlapped by the encephalocele. The right-medial socket was small and shallow and it contained a tiny, dysplastic globe. The left medial socket was better formed and empty.[5]

Facial duplication seems to consist of a spectrum extending from a complete duplication of two faces on the same head to simple additional eye/orbit cavity duplication. There is evidence of right facial nerve palsy noted with abnormal extension of the hairline to the left temporal region.

As previously mentioned, there is a predominance of females over males.[4] However, the case presented here is a male.

Most frequent associated anomalies are anencephaly, duplication of cerebral hemispheres, craniorachischisis, oral clefts, spinal abnormalities, congenital heart defects, diaphragmatic hernia, and thoracic and/or abdominal visceral laterality anomalies.[8] However, this patient presented with posterior plagiocephaly, right facial nerve palsy, and an arachnoid cyst after a through medical examination and investigations were conducted.

Other associated abnormalities such as hypoplasia of the medial temporal lobe, spinal abnormalities such as duplication of the cervical spine, with abnormal cervical and thoracic vertebrae, have been seen.[4] Defects in other organs include diaphragmatic hernia, cardiac defects such as ventricular septal defect, an overriding aorta, a hypoplastic ascending and descending aorta an aortic arch, and dextrocardia. In addition, there may be bilateral dysplastic cystic kidneys, hypoplasia of the ureters, and the urinary bladder, cleft lip, palate, and an imperforate anus.[3] This case has a depressed occipital bone with an abnormally shaped temporal bone. The cardiac, respiratory, gastrointestinal, and genitourinary systems appeared normal on examination.

Due to advances and availability of perinatal imaging modalities, recognition of congenital anomalies *in utero* is possible with the use of 4 D ultrasound scanning machines that give very good definition of facial structures. However, the availability of these diagnostic tools is mostly in tertiary institutions. The index case attended antenatal care in a rural hospital with no such facility thus had no opportunity to have an ultrasound scan done. Prenatal diagnosis of these conditions provides time for early counseling and planning for perinatal management options.

The prognosis is poor for the infants with a complete duplication, although the treatment options such as excision of the duplicated parts, which give a normal appearance in partial diprosopus, have been variably successful. The surgical management of a complex craniofacial malformation such as diprosopus needs a precise morphological analysis of the patient's deformity followed by a clear treatment plan. A staged reconstructive approach is carried out to coincide with facial growth patterns and eye function.[10]

In conclusion, a rare case of a "three-eyed infant" and a review of the literature are presented. The value of perinatal diagnosis and maternal counseling was shown.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

## REFERENCES


1. Bidondo MP, Groisman B, Tardivo A, Tomasoni F, Tejeiro V, Camacho I, *et al*. Diprosopus: Systematic review and report of two cases. Birth Defects Res A Clin Mol Teratol 2016;106:993-1007.
2. Al Muti Zaitoun A, Chang J, Booker M. Diprosopus (partially duplicated head) associated with anencephaly: A case report. Pathol Res Pract 1999;195:45-50.
3. Brugmann SA, Helms JA. Craniofacial formation and congenital defects. In: Sally AM, editor. Principles of Developmental Genetics. Ch. 30. Burlington, MA: Academic Press, an Imprint of Elsevier Inc.;2014.
4. Pradeep S, Mandar D, Nitin V, Vivek M, Sandhya M. Craniofacial duplication: A Case Report. J Clin Diagn Res 2013;9:2025-6.
5. Barr M Jr. Facial duplication: Case, review, and embryogenesis. Teratology 1982;25:153-9.
6. Walton MT, Fineman RM, Walton PJ. Of monsters and prodigies: The interpretation of birth defects in the sixteenth century. Am J Med Genet 1993;47:7-13.
7. Hähnel S, Schramm P, Hassfeld S, Steiner HH, Seitz A. Craniofacial duplication (diprosopus): CT, MR imaging, and MR angiography findings case report. Radiology 2003;226:210-3.
8. Singh S, Chukwunyere DN, Omembelede J, Onankpa B. Foetal congenital anomalies: An experience from a tertiary health institution in North-West Nigeria (2011-2013). Niger Postgrad Med J 2015;22:174-8.
9. Thornton KM, Bennett T, Singh V, Mardis N, Linebarger J, Kilbride H, *et al*. A case of diprosopus: Perinatal counseling and management. Case Rep Pediatr 2014;2014:279815.
10. Kotrikova B, Hassfeld S, Steiner HH, Hähnel S, Krempien R, Mühling J, *et al*. Operative correction and follow-up of craniofacial duplication. Plast Reconstr Surg 2007;119:985-91.