PRENATAL DIAGNOSIS
Prenat Diagn (2009)
Published online in Wiley InterScience
(www.interscience.wiley.com) **DOI:** 10.1002/pd.2321

RESEARCH LETTER

Prenatal 2D and 3D ultrasound diagnosis of diprosopus: case report with post-mortem magnetic resonance images (MRI) and review of the literature

Giuseppe Maria Maruotti[1], Dario Paladini[1], Raffaele Napolitano[1], Laura Letizia Mazzarelli[1], Tiziana Russo[1], Mario Quarantelli[2], Maria Rosaria D'Armiento[3] and Pasquale Martinelli[1]*

[1]*Prenatal Diagnosis Unit, Department of Gynecology and Obstetrics, University Federico II of Naples, Naples, Italy*
[2]*Biostructure and Bioimaging Institute, University Federico II of Naples, Naples, Italy*
[3]*Department of Biomorphological and Functional Sciences, University Federico II of Naples, Naples, Italy*

KEY WORDS: diprosopus; prenatal diagnosis; 2D/3D ultrasonography; magnetic resonance

Conjoined twins represent the rarest type of monozygotic, monochorionic, monoamniotic twins, with an approximate incidence of 1 in 2800 to 1 in 200 000 deliveries. Diprosopus represents an exceedingly rare form of conjoined twins. We present a case of prenatal sonographic characterization of diprosopus in a twin dichorionic pregnancy.

A 26-year-old Caucasian nulliparous woman was referred to our Unit at 22 weeks' gestation of her first pregnancy because of abnormal findings at a second trimester routine ultrasound scan in one twin of a dichorionic pregnancy. The family history was unremarkable. At ultrasound examination we confirmed the dichorionicity of the pregnancy and the normal anatomy of one twin. The other twin had multiple severe anomalies. In particular, two-dimensional (2D) and 3D ultrasound showed anencephaly, four orbits with four eyebulbs and lenses, two noses, micrognatia with both hypoplastic mandibles (Figure 1A and B). The internal organs showed no duplication. Other anomalies included complete rachischisis with scoliosis, transposition of the great arteries, bilateral talipes, and single umbilical artery.

Karyotyping by amniocentesis was proposed but it was declined by the patient. During the counseling, the couple was informed about the type of malformation and decided to continue the pregnancy to term. Premature labor occurred at 37 weeks' gestation and led to a Caesarean section. At birth, the normal male twin had a weight of 2300 g and showed no abnormalities. The abnormal co-twin weighed 1500 g and died few minutes post-partum. On external examination, the external genitalia appeared ambiguous with undescended testes and micropenis. The parents refused autopsy but gross examination confirmed the presence of anencephaly, rachischisis, and facial duplication (four orbits, four eyes, two noses, and two mouths). In addition, there were two external ears, two mandibles, fused adjacent maxillae, and two oral cavities with two fused oral openings. There was a single trunk with a short neck. The limbs appeared normal but bilateral clubfeet were present. No other abnormalities were evident with the exception of fixed contractures of the four limbs.

The parents consented to post-mortem magnetic resonance imaging (MRI), which was performed at 3 Tesla (Magnetom Trio, Siemens medical Systems, Eirlangen, Germany). T2-weighted turbo spin-echo (TSE) images (TR 16 000 ms, TE 210 ms, pixel size 0.5×0.5 mm, slice thickness 2 mm) were acquired along axial, coronal, and sagittal planes. On MRI, in addition to the details detected at ultrasound, three hemimandibles, complete duplication of the nasal bones (Figure 1C), and duplication of the cervical spine with complete schisis extending to the single thoracolumbar tract were demonstrated.

Conjoined twins are the result of incomplete separation of a single ovum, and, as such, the pregnancy is by definition monochorionic. The most accepted theory for their formation is incomplete splitting of a single embryo after the 13th day but before the 25th day after conception (Strauss *et al*., 1987).

The most common type of conjoined twins is thoracopagus (32.7%), with joining at or near the sternal wall and contained viscera, and the rarest type is diprosopus (0.4%), with two faces, one head, and one body. This exceedingly rare variant of conjoined twinning may show a phenotype comprising a wide spectrum and ranging from partial duplication of a few facial structures to complete dicephalus. In the mildest forms, isolated duplication of the nose, eyes, or mouth occurs. The term *tetraophthalmus* is used to describe a fetus with four eyes; in such cases the two median eyes may be partially (Angtuaco *et al*., 1999) or completely fused with a central orbit (Okazaki *et al*., 1987) or may be completely separated, as in our case (Figure 1). In the most severe form of diprosopus, the fetus shows two complete faces.

Diprosopus is mainly found in singletons, but it can occur also in twins, the index case being the third

*Correspondence to: Pasquale Martinelli, Vico Acitillo, 53, 80131, Naples, Italy. E-mail: martinel@unina.it

Received: 5 May 2009
Revised: 22 May 2009
Accepted: 22 May 2009

reported in a dichorionic pregnancy with a normal co-twin, after the reports by Rai *et al*. and Dhaifalah *et al*. In this regard, considering the prevalence of diprosopus in singletons, we agree with Rai *et al*. who raised the possible role of an abnormal twinning process in the etiology of this exceedingly rare condition.

An association with other severe abnormalities has also been reported. These include cleft lip and palate (Chervenak *et al*., 1985) or cleft face (Bulbul *et al*., 2004), neural tube defects (Rydnert *et al*., 1985; Fontanarosa *et al*., 1992; Bulbul *et al*., 2004) (anencephaly, myelomeningocele), heart anomalies, gastrointestinal malformations (gastroschisis, intestinal malrotation, and Meckel's diverticulum), and diaphragmatic hernia. (Angtuaco *et al*., 1999; Fontanarosa *et al*., 1992)

Although prenatal diagnosis of diprosopus has been reported in few cases, (Chervenak *et al*., 1985; Bulbul *et al*., 2004; Rydnert *et al*., 1985; Fontanarosa *et al*., 1992; Angtuaco *et al*., 1999; Okazaki *et al*., 1987; Strauss *et al*., 1987; Rai *et al*., 1998; Picaud *et al*., 1990; Dhaifalah *et al*., 2008) this was only based on 2D imaging. In the present case, this rare condition has been evaluated for the first time with prenatal 3D ultrasound and post-mortem MRI. The recognition of the completely distorted facial anatomy is easily carried out on 2D imaging (Figure 1A). However, if 2D diagnosis of diprosopus is relatively straightforward, considering the easy recognition of the multiple orbits, less simple is, in our opinion, the detailed characterization of the single abnormalities. In our case, 3D ultrasound was helpful to confirm the presence of the four orbits and to study the abnormal bone anatomy on multiplanar imaging. In addition, surface-rendering reconstruction of the diprosopus was able to clearly demonstrate the severe distortion of the fetal facial features and in other case it could help the plastic surgeon in the planning of the future interventions.

Conjoined twins commonly result in early intrauterine demise. Chances of survival and mode of delivery depend on the degree of fusion of the organs and the possibilities of surgical separation. Obstetric care of women with conjoined twins does not differ from that of a normal twin pregnancy, but the newborns require special medical care after delivery. In the rare case of a liveborn with diprosopus, CT, MRI, and MR angiography may provide information about the degree of duplication and to assess if cosmetic correction may be feasible in less severe cases. Post-mortem MRI can add to the characterization of the craniofacial and/or spinal defects. In our case, post-mortem MRI showed duplication of the spine with complete rachischisis and helped us confirm some prenatal sign detected and better characterize other anomalies not visible at ultrasonography, such as the three hemimandibles. MRI also showed complete duplication of the nasal skeleton (Figure 1C). We are aware that such detailed anatomy characterization is probably of very limited clinical relevance, considering the extremely high mortality and the impossibility of a surgical reconstruction in most cases. However, it should be considered when duplication is limited to specific structures, such as the nose (diprosopus dirrhinus), the

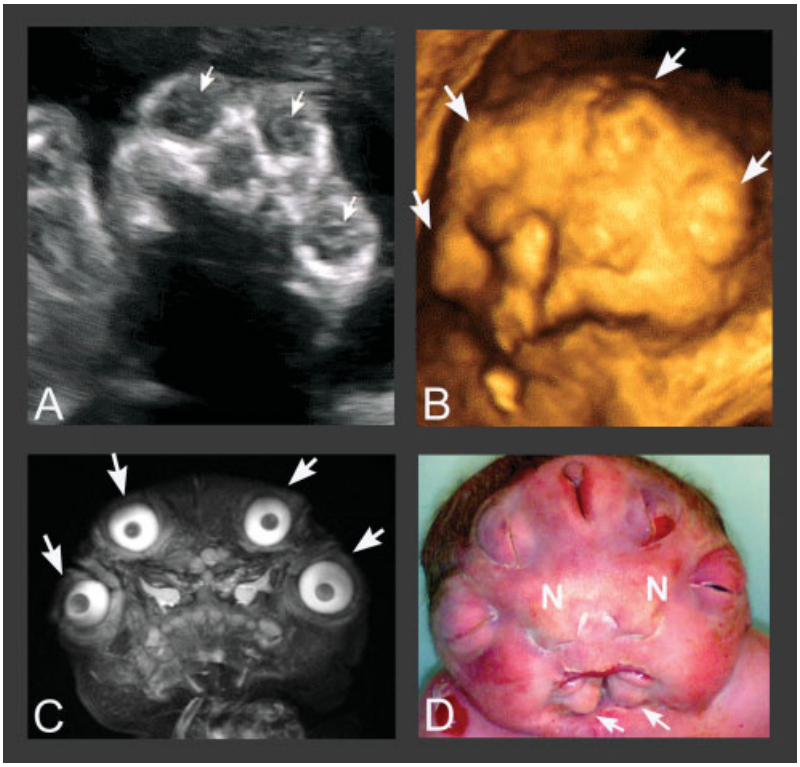

Figure 1—Partial facial duplication in diprosopus as demonstrated at 22 weeks of gestation by two-dimensional (A) and three-dimensional ultrasound (B), post-mortem by magnetic resonance imaging (MRI) (C), and at birth (D). Note the four orbits (arrows), the two noses (N), and the two fused buccal orifices (arrows in D)

DOI: 10.1002/pd

eyes (diprosopus tetraophthalmus), or the mouth only, thus allowing surgical correction. In these extremely rare cases, the surgical management of the complex craniofacial defects needs a precise anatomic analysis of the patient's deformity. 2D and 3D prenatal ultrasound (Figure 1A and B) may be employed in those very rare cases in which the occurrence of only partial fusion/duplication may lead the parents to consider postnatal treatment.

Prenatal MRI may be considered a useful complementary tool in the imaging work-up of fetal cranial and facial structural anomalies. An appropriate imaging strategy is a fundamental part of prenatal diagnosis to allow correct prognostic assessment and postnatal surgical treatment planning.

REFERENCES

Angtuaco TL, Angtuaco EJ, Quirk JG Jr. 1999. US case of the day. Complete brain duplication with fusion at the posterior fossa (diprosopus tetraophthalmos). *Radiographics* **19**(1): 260–263.

Bulbul Y, Drummond CL, Hillion Y, Bidat L, Ville Y. 2004. Diprosopus associated with neural tube defect and facial cleft in the first trimester. *Fetal Diagn Ther* **19**(3): 246–250.

Chervenak FA, Pinto MM, Heller CI, Norooz H. 1985. Obstetric significance of fetal craniofacial duplication. A case report. *J Reprod Med* **30**: 74–76.

Dhaifalah I, Curtisova V, Santavy J. 2008. Prenatal diagnosis of monocephalic bifacial tetraophthalmic diprosopus (conjoined twin). *Fetal Diagn Ther* **23**(1): 82–86, Epub 2007 Oct 9.

Fontanarosa M, Bagnoli G, Ciolini P, Spinelli G, Curiel P. 1992. First trimester sonographic diagnosis of diprosopus twins with craniorachischisis. *J Clin Ultrasound* **20**(1): 69–71.

Okazaki JR, Wilson JL, Holmes SM, Vandermark LL. 1987. Diprosopus: diagnosis in utero. *AJR Am J Roentgenol* **149**(1): 147–148.

Picaud A, Nlome-Nze AR, Engongha-Beka T, Ogowet-Igumu N. 1990. Early prenatal diagnosis of diprosopic syncephalic joined twins. *Rev Fr Gynecol Obstet* **85**(6): 379–381.

Rai VS, Gaffney G, Manning N, Pirrone PG, Chamberlain PF. 1998. Antenatal diagnosis of complete facial duplication—a case report of a rare craniofacial defect. *Prenat Diagn* **18**(6): 618–620.

Rydnert J, Holmgren G, Nielsen K, Bergman F, Joelsson I. 1985. Prenatal diagnosis of conjoined twins (diprosopus) with myelomeningocele. *Acta Obstet Gynecol Scand* **64**: 687–689.

Strauss S, Tamarkin M, Engelberg S, Ben Ami T, Goodman RM. 1987. Prenatal sonographic appearance of diprosopus. *J Ultrasound Med* **6**: 93–95.

DOI: 10.1002/pd