

True Nodes of the Umbilical Cords of a Mono-Amniotic Pregnancy

Astou Coly Niassy*, Ibrahim Rahadat, Aissatou Sankhare, Marie-Edouard Faye

Gynecology-Obstetrics Hospital Aristide Le Dantec, Dakar, Senegal

Email: *medacn@gmail.com

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Abstract

The incidence of twin pregnancies, has increased due to assisted reproductive technologies [1]. We note a doubling between 1970 and 2010 from less than 8‰ to nearly 16‰ of pregnancies (Pison *et al.*, 2014; Pison *et al.*, 2015) in developed countries [2] and 17‰ to 20‰ in sub-Saharan Africa. Monozygotic twins are the product of fertilization of a single egg by a single sperm, with the egg splitting into two within the first few days after fertilization. If the division takes place after the 8th day, the pregnancy is mono-chorionic mono-amniotic (less 1% of cases of monozygotic pregnancies) and represents 1 in 30,000 pregnancies. It is the development of two fetuses within the same amniotic sac, representing the rarest type of twin pregnancy (1% of monozygotic twin pregnancies) but being the one with the highest fetal and maternal morbidity. The main complication feared is coiling and knots of the umbilical cords responsible for abnormal heartbeat, fetal suffering and eventual fetal death. We report the case of in utero fetal death of twins from a mono chorionic mono amniotic pregnancy secondary to knots and coiling of the cord in order to discuss the diagnostic and therapeutic aspects.

Keywords

Twin Pregnancy, Fetal Death, Coiling

1. Introduction

Twin pregnancy once rare, has seen an increase in its incidence worldwide by the advent of medically assisted reproduction techniques [1] with a doubling of its incidence between 1970 and 2010 from less than 8‰ to nearly 16‰ in developed countries [2]. In sub-Saharan Africa its incidence seems to be increasing with an average of 17 and 20 twin deliveries per thousand (‰) total deliveries [2]. We distinguish monozygotic (MZ) or identical twins and dizygotic (DZ) or fraternal

twins. Monozygotic twins are the product of the fertilization of a single egg by a single spermatozoon, the egg splitting into two in the first few days after fertilization. They can be bichorial-bi amniotic representing 30% of the monozygotic pregnancies (1/1000 pregnancies) if the division of the fertilized egg takes place between 1 and the 5th day, mono chorial bi amniotic the most frequent, it represents 70% of monozygotic pregnancies (2/1000 pregnancies) if the division took place between the 5th and the 8th day, after the 8th day the pregnancy is mono chorial mono amniotic (less 1% of monozygotic pregnancies) and represents 1 for 30,000 pregnancies. Dizygotic pregnancies are always bi-chorionic and bi-amniotic [3].

Problems posed: problem of early diagnosis of monoamniotic pregnancy.

Does it require a particular surveillance for its patients, *i.e.* a hospitalization in the third trimester for a better surveillance of an anomaly of the fetal cardiac rhythm?

2. Patient and Observation

It is about Mrs M.C aged 19 years, primigravida primiparous with family history of twin pregnancy married under monogamous regime without notion of taking ovulation inducer which was referred to us for a management of a fetal death in utero of the twins on mono choriale monoamniotic twin pregnancy. The pregnancy was poorly monitored with a late prenatal consultation and ultrasound performed in the second trimester of pregnancy which showed a monoamniotic twin pregnancy of 24 SA.

She received a dose of sulfadoxine-pyrimethamine and was supplemented with iron and folic acid.

The clinical examination revealed a blood pressure of 12/8 cmHg, a pulse of 110 bpm, slightly colored mucous membranes, and soft calves.

The obstetrical examination showed a uterine height of 42 cm. The patient could not feel the fetuses moving. The fetal heart sounds were not perceived with the obstetrical stethoscope and the vaginal touch found a long posterior cervix admitting a finger with intact membranes and first twin in cephalic presentation.

The ultrasound scan showed a monoamniotic twin pregnancy stopped at 32 weeks of amenorrhea.

The maternal check-up showed a normocytic normochromic anaemia at 7 g/dl, the blood count was normal.

In total we concluded to a fetal death in utero on twin pregnancy not in labor with an unfavorable bishop score associated with anemia. A delivery by caesarean section was recommended with evidence of umbilical cord knots and a snowshoe insertion at the placental level as shown in **Figure 1** and **Figure 2**.

Both fetuses were male. Ultrasound confirmed the diagnosis of fetal death, but the decrease in amniotic fluid did not allow proper study of the umbilical cords (**Figure 3**).

3. Discussion

Mono-amniotic pregnancies are extremely rare. Each twin has its own umbilical



Figure 1. Double knot of umbilical cords.



Figure 2. Snowshoe insertion.



Figure 3. Decrease in amniotic fluid.

cord. Wharton's jelly, which gives the umbilical cord its flexibility and elasticity, protects the umbilical vessels against the effects of false knots and moderate compression [1] [2].

The true knot, on the other hand, is responsible for serious fetal complications: in utero growth retardation, per partum asphyxia, and fetal death in utero. It leads to partial or complete obstruction of fetal blood flow.

The incidence of true umbilical cord knot is between 0.3% and 1.3% of pregnancies.

The problem posed is that of early diagnosis of monoamniotic pregnancy, making it possible to anticipate this complication which remains unpredictable [3] [4].

Mono amniotic twins are a rare but important subset of twins at risk for unique and serious complications, placing them at the highest risk of perinatal mortality of all twin gestations. In addition to the risks faced by all twins (prematurity, selective growth retardation), all mono chorionic twins (transfusion-transfusion syndrome), and all monozygotic twins (congenital anomalies), mono amniotic twins face the unique risk of cord entanglement. Therefore, early diagnosis, screening for fetal anomalies, monitoring for transfusion-transfusion syndrome, decisions related to post-viability monitoring, and timing and route of delivery are all essential. There are no recommendations for inpatient or outpatient management, timing of delivery, and route of delivery. A study called the MONOMONO [3] [4] study was done in 22 university hospitals in Italy, the United States, the United Kingdom and Spain, from January 2010 to January 2017 with the aim of assessing fetal morbidity and mortality according to gestational age in mono amniotic twins from 26 weeks of gestation and comparing outpatient to inpatient monitoring and showed that in uncomplicated mono amniotic twins, in-hospital monitoring was associated with similar fetal mortality to outpatient management after 31 + 6 weeks, and up to 36 + 6 weeks, there were no intrauterine fetal deaths or neonatal deaths. A retrospective analysis of all mono amniotic pregnancies managed at Karolinska University Hospital, Stockholm, Sweden 2010-2019 was performed all uncomplicated mono amniotic twin pregnancies are followed as outpatients and delivered by elective cesarean section in gestation weeks 32 - 34.

A multinational cohort study by Saccone *et al.* evaluated the perinatal mortality rate of patients managed as inpatients versus outpatients in uncomplicated monoamniotic twin pregnancies. The results suggest that the perinatal mortality rate was lower in the inpatient group, but the results were not conclusive [3] [4].

Caesarean section between 33 and 34 weeks, results in a substantial reduction in neonatal morbidity.

The risk of an unpredictable funicular accident and its other specific complications requires special surveillance and systematic cesarean section before 36 weeks (CNGOF). Some teams request hospitalization of these patients at risk for better monitoring. A case of multiple and complex umbilical cord knots discov-

ered during caesarean section with extraction of live newborns with an Apgar score of 7 has been reported [5] [6].

Does it require special monitoring for these patients, *i.e.* hospitalization in the third trimester for better monitoring of fetal heart rate abnormality? The length of the cord can be estimated by studying the loops, but it is impossible to give an exact measurement. Factors that may contribute to cord knots include: abnormally long cord length, excess amniotic fluid (hydramnios), small fetus, mono-chorionic/monoamniotic pregnancy, as in our case. Male fetuses have an increased risk of cord coiling with knots, with cord length differing between girls and boys after 27 weeks of pregnancy. The greater length of the umbilical cord in boys, as well as their greater physical activity, may explain the greater frequency of knots and strangulations.

Studies have pointed out that multiparity is also a risk factor.

When the knot is tight, the risk of fetal death is increased fourfold. Fortunately, the outcomes are not always fatal. In 2003, American doctors (Boston, USA) reported the case of a 29-year-old woman who delivered a 3.7 kg child born at term by vaginal delivery [7].

Similarly, in 2005, a British team described a woman with a monoamniotic pregnancy. An emergency caesarean section was performed at 33 weeks of pregnancy due to the occurrence of fetal distress. Despite the complexity of the knot, both babies were doing well.

Finally, more recently, in April 2020, Iranian obstetricians reported the case of a newborn, extracted by cesarean section, whose umbilical cord measured 70 cm and had not one but two knots [8] [9] [10]. The physicians observed no deleterious consequences on the development of the child. His Apgar score was 9 at one minute and 10 at five minutes. Indeed, in two other similar cases, published in 1998 and 2017, both cord knots had resulted in intrauterine growth restriction (IUGR) [6] [7] [11].

4. Conclusion

A rare but serious complication, the true cord knot can engage the vital or functional fetal prognosis. The surveillance in case of pregnancy at risk must be codified and adapted to the context.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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