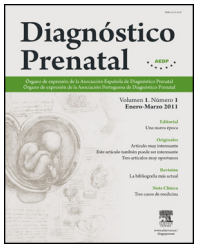
DIAGN PRENAT. 2013;xxx(xx):xxx–xxx

Diagnóstico Prenatal

www.elsevier.es/diagnprenat



Original Article

Neurological morbidity of monochorionic twins

Isaac Blickstein[a,b]

[a] *Department of Obstetrics and Gynecology, Kaplan Medical Center, Rehovot, Israel*
[b] *The Hadassah-Hebrew University School of Medicine, Jerusalem, Israel*

ARTICLE INFO

Article history:
Received 31 July 2013
Accepted 28 August 2013
Available online xxx

Keywords:
Monochorionic twins
Neurological morbidity
Porencepahly
Cerebral palsy

ABSTRACT

Monochorionic twins (MC) are at increased risk for morbidity. The unique placenta with vascular anastomoses may create imbalance with either acute or chronic hypotension or cardiac insufficiency that will eventually affect the fetal brain. It appears that these characteristics of the MC placenta add to the already higher frequency of brain anomalies observed among MZ twins.

TOPS, TAPS, and TTTS require not only inter-twin anastomoses but also two live twins with an unbalanced shunt of blood. The death of the co-twin may prompt a sudden hypotension in the surviving twin with the consequent brain lesions. The more frequently occurring velamentous cord insertion in this kind of pregnancies is related to severe selective intrauterine growth restriction that may cause cerebral compromise. Finally, the preterm birth rate among MC twins is ten times higher than in singletons, and prematurity is a key factor for neurological morbidity as well.

In this paper the various aspects of neurological morbidity in MC twins will be discussed.

Morbilidad neurológica en gemelos monocoriónicos

Palabras clave:
Gemelos monocoriónicos
Morbilidad neurológica
Porencefalia
Parálisis cerebral

RESUMEN

Los gemelos monocoriónicos (MC) poseen mayor riesgo de presentar un mal pronóstico, especialmente en la morbilidad neurológica. La existencia de una única placenta con anastomosis vascular crea un desequilibrio hemodinámico que causa hipotensión aguda o crónica, o insuficiencia cardiaca, lo que eventualmente afectará al cerebro fetal. Parece que estas características de la placenta MC se suman a la ya mayor frecuencia de anomalías cerebrales observadas entre los gemelos MZ.

La secuencia oligodramnio y polidramnio (TOPS), las secuencias anemia-policitemia (SAP) y el síndrome de transfusión fetal-fetal (STFF) requieren no solo anastomosis intergemelar, sino que además deben existir 2 gemelos vivos con un desequilibrio en el intercambio sanguíneo. La muerte de uno de los cogemelos puede provocar una hipotensión brusca en el gemelo superviviente con las consiguientes lesiones cerebrales. La inserción velamentosa del cordón, que ocurre con mayor frecuencia en este tipo de embarazos, está relacionada con una restricción grave del crecimiento intrauterino selectivo que puede comprometer

E-mail address: blick@netvision.net.il

2173-4127/$ – see front matter
http://dx.doi.org/10.1016/j.diapre.2013.08.001

Please cite this article in press as: Blickstein I. Neurological morbidity of monochorionic twins. Diagn Prenat. 2013. http://dx.doi.org/10.1016/j.diapre.2013.08.001

la función cerebral. Por último, la tasa de nacimientos prematuros entre gemelos MC es 10 veces superior a la de los embarazos simple s, y la prematuridad también es un factor clave para la morbilidad neurológica.

En este artículo se examinarán los distintos aspectos de la morbilidad neurológica en los gemelos MC.

Introduction

Zygotic splitting is a formidable embryological event whereby an embryo that would otherwise develop into a singleton undergoes some unknown changes that lead to an embryological accident. This accident splits the early embryo and is responsible for the numerous malformations seen in these so-called monozygotic (MZ) twins.[1] It is assumed (never proven) that if this embryological event occurs soon after fertilization, the placenta also splits and the pregnancy becomes a bichorionic (BC) twin gestation. However, when the insult is somewhat delayed, the placenta is not spared and the resultant monochorionic (MC) pregnancy demonstrates severe placental malformations never seen in any other pregnancy.

The placental malformations include 3 elements. First, the placental territory that supplies each fetus is rarely equal. That said, unequal or discordant placental sharing is the rule rather than the exception, and some degree of discordant growth related to the unequal placental territory is common. In severe disproportion between the placental shares, selective intrauterine growth restriction (sIUGR) develops.

The second placental malformation in MC twins is the existence of inter-twin vascular connections (anastomoses). These come in various forms (veno-venous, arterio-arterial, arterio-venous) and calibers. In a very simplified version, this construct might lead to an imbalance shift of blood (transfusion) between the two twins involving deep arterio-venous anastomoses. At the end, a net twin–twin blood transfusion may occur (TTTS) which initiates a series of cardiac events in the overloaded recipient which develops polyhydramnios, hormonal messages from the recipient to the hyppovolemic donor which develops oligohydramnios, forming the twin oligo-polyhydramnion sequence (TOPS)—the first stage and hallmark of TTTS. Major changes in the definition, diagnosis and treatment of TTTS were observed in the last 25 years. Primarily,[2] the diagnosis changed from a postnatal to an antenatal diagnosis. Next, better understanding of the pathogenesis as well as improved imaging led to establishing new stages of TTTS and, finally, various treatment modalities were examined. TTTS is a serious complication, and if remained untreated, may lead to single or double deaths.

The intertwin anastomoses—invariably present in the MC placenta—may also cause discordant hemoglobin levels once believed to be a criterion for TTTS. This anemia–polycythemia sequence (TAPS) may be seen with or without TTTS, and not infrequently after laser photocoagulation treatment of TTTS.

Transfusion through intertwin anastomoses does not necessarily have a bad connotation. For example, some small twins in the setting of TTTS survive in utero only because the larger twin supplies its growth restricted twin by a A–A anastomosis (the so-called 'rescue' anasthomosis).

TOPS, TAPS, and TTTS require not only inter-twin anastomoses but also two live twins. In the case of single fetal demise it was once believed that the dead twin may transfuse thromboplastin-like emboli through the vascular connections, leading to end-organ damage in the survivor. This mechanism was termed twin embolization syndrome irrespective of the fact that emboli were not found. Further research found that instead of embolization from the dead twin to the survivor, the shift of blood is from the survivor (normal blood pressure) to the dead (low blood pressure) twin via the anastomoses. In this scenario, the loss of blood may cause death of the survivor soon after its dead co-twin, or in less significant blood loss, hypovolemic damage to susceptible organs like the brain, kidney, adrenals, etc. Minor blood loss would result in an intact survivor.

The third placental malformation is the pathological insertion of the umbilical cord, usually to the placental side—the so-called velamentous cord insertion. It appears that this malformation is associated with both TTTS and sIUGR.

It appears that these characteristics of the MC placenta add to the already higher frequency of brain anomalies observed among MZ twins. Nevertheless, the major neurological threat to a MC pregnancy is not only being a twin pregnancy with an inherent higher risk of prematurity compared to singletons but also being a MC twin pregenancy with an inherent higher risk of prematurity compared to DC twins.

In this paper the various aspects of neurological morbidity in MC twins will be discussed.

Being a twin

Twinning is associated with increased risk of cerebral palsy (CP) with an average prevalence of 7.4% twins among CP cases. The prevalence of CP was roughly 6 times higher than that in singletons.[3] It goes without saying that the most significant contributor to this increased rate is over-representation of twins among low and very low birth weight (LBW/VLBW) and among preterm and very preterm infants.

However, when stratifying the prevalence of CP in twins and singletons according to birth weight and gestational age, the data suggest that multiple and singleton pregnancies have similar risks for CP until around 36–37 weeks. It follows that although LBW/VLBW and preterm birth are the most significant risk factors for CP, the disadvantage of twins is apparent near term when the risk for singletons is extremely low. This conclusion may imply that 'term' occurs earlier in twins, and supports the recommendation to deliver all twins by 38 weeks gestation.

Please cite this article in press as: Blickstein I. Neurological morbidity of monochorionic twins. Diagn Prenat. 2013. http://dx.doi.org/10.1016/j.diapre.2013.08.001