Journal of Regenerative Biology and Medicine

ISSN: 2582-385X
Bittmann S. 2021- J Regen Bio Med
Case Report

One of Curious Congenital Midline Abdominal Defects In Newborn: A Giant Omphalozele In A 3 Years-Old Boy

Stefan Bittmann, Gloria Villalon, Elena Moschüring-Alieva, Elisabeth Luchter

*These authors contributed equally to this work

Department of Pediatrics, Ped Mind Institute (PMI), Gronau, Germany

*Corresponding Author: Stefan Bittmann, Head of the Department of Pediatrics and Ped Mind Institute (PMI) Pediatrician, Hindenburgring, Gronau, Germany.

Accepted Date: 03-05-2021
Published Date: 03-06-2021

Introduction

Omphalocele is one of the congenital abdominal wall defects and specifically refers to an umbilical ring defect in which the abdominal organs protrude in front of the abdominal wall in a thin sac or membrane. Such an abdominal wall defect develops around the third week of pregnancy due to the failure of adhesion between the embryonic lateral abdominal wall components. The condition occurs in 1 in 5,000 children, with boys more commonly affected than girls [1,2,3,9].Overall, it is a rare disease whose therapy requires the experience of proven specialists [1-9,13-17]. For this purpose, pediatric surgeons are available to assist children and their parents from the time of diagnosis in the womb through direct care in the delivery room and, if necessary, further surgery to follow-up treatment. Through close and direct contact with the individual physicians, one can achieve an optimal and most comfortable care situation for the child. During embryonic development, part of the intestine emerges from the abdominal cavity of the embryo into the amniotic cavity to turn around. As the pregnancy progresses, the intestine return to the abdomen of the child and the abdominal wall closes. If the abdominal wall does not close or closes incompletely, an abdominal wall defect results and parts of the abdominal organs (mostly the intestine) are located further outside the abdominal cavity. In some cases the intestine is covered by a membrane of the umbilicus.

Definition of Exomphalos (omphalocele)

Exomphalos is a midline defect with herniation of abdominal contents into the base of the umbilical cord, confined by an amnioperitoneal membrane. In a large exomphalos, liver as well as intestine may be present in the sac. There is a high incidence of chromosomal abnormalities like Trisomy 18 and 13, and the finding of an exomphalos is an indication for fetal karyotyping. Overall, in two-thirds of cases (particularly those with aneuploidy) there are other structural anomalies, often multiple and frequently congenital heart disease and it is a feature of a number of recognisable conditions. All these conditions are rare but somecan present with features that may be detected on ultrasound, and some have a high recurrence risk for subsequent pregnancies.

Midline Abdominal Defects in Childhood

Citation: Bittmann S, Villalon G, Moschüring-Alieva E, Luchter E. One of Curious Congenital Midline Abdominal Defects In Newborn: A Giant Omphalozele In A 3 Years-Old Boy. J Regen Biol Med. 2021;3(3)1-5.
DOI: https://doi.org/10.37191/Mapsci-2582-385X-3(4)-073

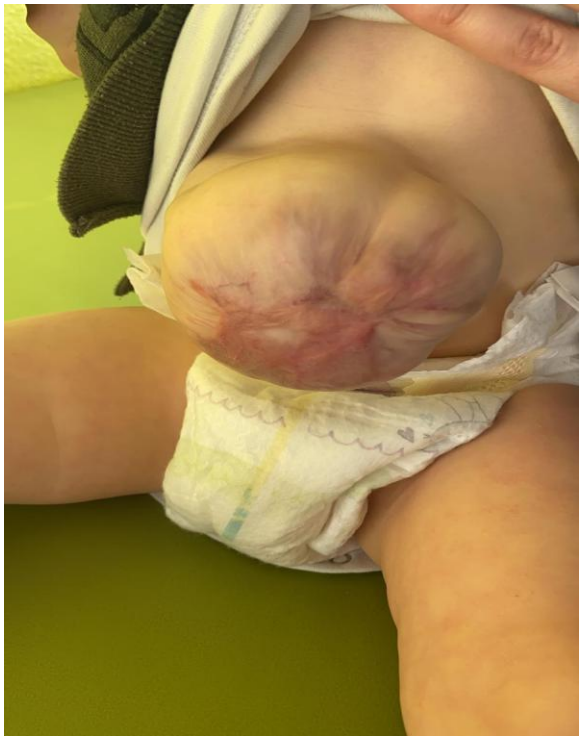
There is a list of 7 different midline abdominal defects:

a) Bladder Exstrophy
b) Body-Stalk Anomaly
c) Cloacal Exstrophy
d) Ectopia Cordis
e) Gastroschisis
f) Omphalocele
g) Pentalogy of Cantrell

Course, Therapy and Follow-up in cases of Omphalozele

The children who are known to have an abdominal wall defect are usually born via cesarean section. While still in the delivery room, the exposed abdominal organs are wrapped sterilely and warmly. Surgical treatment is usually performed on the same day. The type of care depends on the size of the abdominal wall defect and the volume of the exposed abdominal organs. If it is a small defect with only a small amount of intestine protruding, the intestine can be moved back into the abdomen and the abdominal wall can be reconstructed. However, if there is a larger defect with large portions of the abdominal organs outside the abdominal cavity, primary closure of the abdominal wall is usually not successful. In these cases, either a so-called silo bag is inserted as a temporary abdominal wall closure or a plastic membrane is sewn in (Schuster plastic). Within a few days, the abdominal wall will have expanded so that the abdomen can be closed. In many cases, children can drink normally after the abdominal wall is closed. In some cases, however, additional intestinal malformations are present that need to be corrected (intestinal atresia). Rarely, the length of the intestine is also restricted, so that some of the children require special food or even have to be supplied with nutrients via the vein. Treatment is provided by an interdisciplinary team of prenatal diagnosticians, gynecologists & obstetricians, anesthesiologists, neonatologists and pediatric surgeons. If there is an intact hernia sac in the umbilical cord area without signs of perforation or infection, elective care can be performed. For this purpose, the hernia sac is protected by moist cloths and "silo" packs. After clarification of other malformations, surgical treatment is performed. The course depends on the size of the omphalocele and the accompanying malformations. Food buildup is often difficult and several surgeries may be required until complete abdominal closure is achieved. The prognosis depends on the other malformations and not on the omphalocele or necessary number of operations until abdominal wall closure.

Figure 1: Large omphalozele of a 3-years-old boy



Discussion

The incidence of omphalocele is 1:4000 - 1:5000 births [13-15,19]. The omphalocele is not hereditary and usually does not result in a higher risk for further children of the same mother. Ultimately, the cause (etiology) of omphalocele is currently still unclear [1-11,12-14]. In about four out of ten children with an omphalocele, other malformations are present, e.g. in the area of the heart, kidneys, liver and intestines [1-19]. Omphaloceles are found with above-average frequency in children with

Citation: Bittmann S, Villalon G, Moschüring-Alieva E, Luchter E. One of Curious Congenital Midline Abdominal Defects In Newborn: A Giant Omphalozele In A 3 Years-Old Boy. J Regen Biol Med. 2021;3(3)1-5.
DOI: https://doi.org/10.37191/Mapsci-2582-385X-3(4)-073

Cantrell syndrome, Pätau syndrome (trisomy 13), Edwards syndrome (trisomy 18), Fraser syndrome, trisomy 16 and triploidy. Furthermore, they occur in Shprintzen syndrome type I and Beckwith-Wiedemann syndrome [8-12].

The course depends on the size of the omphalocele and any accompanying malformations. In infants, the build-up of food is sometimes difficult, so that food must be fed by tube for a certain period of time, possibly even in infancy. In the case of larger omphaloceles, several operations are often necessary until the abdominal wall is completely closed.

Due to the further development of sonographic examinations for early diagnosis, as well as improved possibilities for surgical treatment and parenteral nutrition, the prognosis for omphaloceles has improved significantly. A 2009 Dutch study summarizes: After intensive medical management early in life, long-term outcomes are similar for both patients with smaller and larger findings for omphalocele, except for cosmetic considerations, which are more severe for large omphaloceles. However, this did not affect the quality of life in either group, which compares favorably with that of healthy young adults. Taking into account this positive outlook, expectant parents with an omphalocele prenatally detected in the child, as well as parents of newborns with it, should be informed that the high burden of (surgical) therapy to which the child must necessarily be subjected is very likely to result in a good state of health in the long term, especially if no other anomalies are present. The quality of life of patients with omphalocele is not significantly different from the population average, according to other studies as well [1-19]. There is no shortened life expectancy or higher prevalence of disease, nor have any differences been found in terms of educational attainment. Pregnancy is possible for female patients.

References

1. Krause H, Pötzsch S, Hass HJ, Gerloff C, Jaekel A, Avenarius S, Kroker S. VentraleBauchwanddefekte--Darstellung der Entwicklung in Prävalenz und operativemVorgehenanhand von Gastroschisis und Omphalozele [Congenital abdominal wall defects--an analysis of prevalence and operative management by means of gastroschisis and omphalocele]. ZentralblChir. (2009) Dec;134(6):524-31. German. doi: 10.1055/s-0029-1224681. Epub 2009 Dec 17. PMID: 20020384.
2. Schier F, Schier C, Stute MP, Würtenberger H. 193 Fälle von Gastroschisis und Omphalozele--Postoperative Ergebnisse [193 casesofgastroschisisandomphalocele--postoperative results]. ZentralblChir. (1988) ;113(4):225-34. German. PMID: 2966527.
3. Stratmann C. Gastroschisis und Omphalozele, anästhesiologische Aspekte [Gastroschisisandomphalocele, anesthesiologicaspects]. AnasthesiolIntensivmedNotfallmedSchmerzther (1997) Aug;32(8):513-4. German. doi: 10.1055/s-2007-995102. PMID: 9376469.
4. Binet A, Gelas T, Jochault-Ritz S, Noizet O, Bory JP, Lefebvre F, Belouadah M, James-Robert I, Aubert D, Bouche-PillonPersyn MA, Poli-Merol ML, François-Fiquet C. VAC® therapy a therapeutic alternative in giant omphalocele treatment: a multicenter study. J PlastReconstrAesthet Surg (2013) Dec;66(12):e373-5. doi: 10.1016/j.bjps.2013.05.010. Epub 2013 May 28. PMID: 23721627.
5. van Zalen-Sprock RM, Vugt JM, van Geijn HP. First-trimester sonography of physiological midgut herniation and early diagnosis of omphalocele. Prenat Diagn (1997) Jun;17(6):511-8. PMID: 9203209.

Citation: Bittmann S, Villalon G, Moschüring-Alieva E, Luchter E. One of Curious Congenital Midline Abdominal Defects In Newborn: A Giant Omphalozele In A 3 Years-Old Boy. J Regen Biol Med. 2021;3(3)1-5.
DOI: https://doi.org/10.37191/Mapsci-2582-385X-3(4)-073

6. Na Q, Liu C, Cui H, Zhang Z, Yin S, Li Q. Immediate repair compared with delayed repair of congenital omphalocele: short-term neonatal outcomes in China. J Int Med Res. (2011) 39(6):2344-51. doi: 10.1177/147323001103900634. PMID: 22289553.
7. Eltayeb AA, Mostafa MM. Topical treatment of major omphalocoele: Acacia nilotica versus povidone-iodine: A randomised controlled study. Afr J Paediatr Surg (2015) Oct-Dec;12(4):241-6. doi: 10.4103/0189-6725.172553. PMID: 26712288; PMCID: PMC4955476.
8. Youssef F, Gorgy A, Arbash G, Puligandla PS, Baird RJ. Flap versus fascial closure for gastroschisis: a systematic review and meta-analysis. J Pediatr Surg (2016) May;51(5):718-25. doi: 10.1016/j.jpedsurg.2016.02.010. Epub 2016 Feb 11. PMID: 26970850.
9. Ha LC, Craig A, Grace MR, Osmundson SS, Taylor EW, Zuckerwise LC. Accuracy of estimated fetal weight assessment in fetuses with abdominal wall defects. Am J Obstet Gynecol MFM. (2021) Apr 22:100385. doi: 10.1016/j.ajogmf.2021.100385. Epub ahead of print. PMID: 33895400.
10. Prefumo F, Izzi C. Fetal abdominal wall defects. Best Pract Res Clin ObstetGynaecol (2014) Apr;28(3):391-402. doi: 10.1016/j.bpobgyn.2013.10.003. Epub 2013 Dec 3. PMID: 24342556.
11. Kelly KB, Ponsky TA. Pediatric abdominal wall defects. Surg Clin North Am. (2013) Oct;93(5):1255-67. doi: 10.1016/j.suc.2013.06.016. Epub 2013 Jul 26. PMID: 24035087.
12. Gamba P, Midrio P. Abdominal wall defects: prenatal diagnosis, newborn management, and long-term outcomes. Semin Pediatr Surg. (2014) Oct;23(5):283-90. doi: 10.1053/j.sempedsurg.2014.09.009. Epub 2014 Sep 4. PMID: 25459013.
13. Ledbetter DJ. Congenital abdominal wall defects and reconstruction in pediatric surgery: gastroschisis and omphalocele. Surg Clin North Am. (2012) Jun;92(3):713-27, x. doi: 10.1016/j.suc.2012.03.010. Epub 2012 Apr 17. PMID: 22595717.
14. Islam S. Advances in surgery for abdominal wall defects: gastroschisis and omphalocele. ClinPerinatol (2012) Jun;39(2):375-86. doi: 10.1016/j.clp.2012.04.008. PMID: 22682386.
15. Youssef F, Gorgy A, Arbash G, Puligandla PS, Baird RJ. Flap versus fascial closure for gastroschisis: a systematic review and meta-analysis. J Pediatr Surg (2016) May;51(5):718-25. doi: 10.1016/j.jpedsurg.2016.02.010. Epub 2016 Feb 11. PMID: 26970850.

16. Lakshminarayanan B, Lakhoo K. Abdominal wall defects. Early Hum Dev. 2014 Dec;90(12):917-20. doi: 10.1016/j.earlhumdev.2014.09.018. Epub (2014) Oct 14. PMID: 25448781.
17. Tassin M, Benachi A. Diagnosis of abdominal wall defects in the first trimester. CurrOpinObstet Gynecol (2014) Apr;26(2):104-9. doi: 10.1097/GCO.0000000000000053. PMID: 24504173.
18. Victoria T, Andronikou S, Bowen D, Laje P, Weiss DA, et al. Fetal anterior abdominal wall defects: prenatal imaging by magnetic resonance imaging. Pediatr Radiol (2018) Apr;48(4):499-512. doi: 10.1007/s00247-017-3914-x. Epub 2018 Mar 17. PMID: 29550866.
19. Bittmann S, Ulus H, Springer A. Combined Pentalogy of Cantrell with tetralogy of Fallot, gallbladder agenesis, and polysplenia: a case report. J Pediatr Surg (2004) Jan;39(1):107-9. doi: 10.1016/j.jpedsurg.2003.09.030. PMID: 14694384.