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The relation between viscero-abdominal disproportion and type of omphalocele closure

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ABSTRACT

Objective: To investigate the relation between prenatal ultrasound measurements of viscero-abdominal disproportion and the expected type of postnatal surgical closure of an omphalocele.
Study design: Retrospectively, 24 fetuses diagnosed with an isolated omphalocele in the 2nd trimester of pregnancy were selected (period 2003–2013). An image of the axial plane of the abdomen at the level of the defect was retrieved. The ratio of omphalocele circumference to abdominal circumference (OC/AC), and the ratio of defect diameter to abdominal diameter (DD/DA) were calculated. Prognostic outcome was primary closure. Sensitivity and specificity and the corresponding area under the ROC curve of these ratios were calculated as measurements of prognostic accuracy.
Results: Primary closure was achieved in 15/24 cases. For the OC/AC-ratio a cut-off value of 0.82 successfully predicted outcome in 23/24 cases with an area under the ROC curve of 0.99. A cut-off value of 0.61 for the DD/DA-ratio successfully predicted type of closure in 20/24 cases with an area under the ROC curve of 0.88. In all cases without eviscerated liver tissue, the defect was primarily closed.
Conclusion: In prenatal isolated omphalocele cases, the OC/AC-ratio is better at predicting postnatal surgical closure than the DD/DA-ratio and can be used as a prognostic tool for expected type of closure in the 2nd trimester of pregnancy.

Introduction

An omphalocele is a congenital defect of the anterior abdominal wall, with a birth prevalence of approximately 1:12.5000 [1]. Depending on the size of the defect, intestines, liver and/or gallbladder are herniated through the defect [2]. Additional congenital anomalies and/or syndromes, with or without abnormal karyotype, are found in approximately 80% of cases [3]. Associated anomalies rather than the omphalocele alone are decisive for treatment decision and determine clinical outcome [4,5]. Depending on gestational age at diagnosis and presence of associated abnormalities, termination of pregnancy (TOP) is performed in up to 83% of cases [1,3,5–7].

In isolated cases postnatal outcome depends on type of surgical closure (primary or delayed) [8]. Primary closure is defined as closure immediately after birth. Delayed closure is defined as late or even post-infancy closure after allowing the omphalocele to desiccate, contract and epithelialize with closure of the ventral hernia at a later stage. Type of surgical closure is determined by the size of the defect, evisceration of the liver [9], intra-abdominal pressure, duration of mechanical ventilation, inspiratory oxygen fraction and clinical presentation of pulmonary hypoplasia [10]. Delayed closure is associated with increased co-morbidity and extended hospital stay [11,12].

The vast majority of the omphaloceles is diagnosed in the first or second trimester of pregnancy. As there are no guidelines on case specific assessment, counseling on individual prognosis is not yet possible [3,7,13–15], Individual counseling, however, enables parents to anticipate for a relative long period of hospitalization of their infant [3,15]. We report on additional ultrasound measurements for omphalocele closure which are relevant to the counseling period, i.e. prior to 24 weeks of gestation.

Materials and methods

This retrospective cohort study was conducted in our tertiary referral center between January 2003 and December 2013. For the study we restricted ourselves to isolated omphaloceles diagnosed

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prior to 24 weeks of gestation. As a result, cases were not selected if they were diagnosed prenatally with major associated anomalies or karyotype with lethal prognosis (i.e. trisomy 13 or 18), which would influence type of postnatal closure. Cases diagnosed postnatal with additional anomalies were not excluded, since this information was unknown at the time of the prenatal diagnosis and counseling. Cases were excluded when parents had decided to terminate the pregnancy, an intra-uterine fetal death (IUFD) had occurred, or infants had died shortly after birth before any attempt of surgical treatment. Also, cases were excluded if no ultrasound images were available depicting the axial plane of the abdomen at the level of the defect for measuring the required ratios.

The primary outcome variable was type of closure (primary or delayed). The indications for the type of surgical closure were the same for all cases, according to the consensus of the Dutch society of pediatric surgery [16]. The following maternal and fetal characteristics of the included cases were obtained: gestational age at ultrasound examination, omphalocele contents, presence of other associated anomalies (detected prenatally or postnatally), fetal karyotype, and type of closure (Table 1). In cases where there was more than one ultrasound examination prior to 24 weeks of gestation we selected the one closest to 20 weeks of gestation.

We studied two ratios as candidate predictors of successful primary closure. The OC/AC-ratio was measured excluding the edema by dividing the omphalocele circumference by the abdominal circumference in the same ultrasound image at the level of the defect (Fig. 1.1). The OC/AC-ratio was measured to assess the size of an omphalocele in comparison to the fetal abdomen to quantify the degree of viscero-abdominal disproportion. This resembles the assessment by pediatric surgeons, who estimate the viscero-abdominal disproportion postnatally to determine whether a defect can be primarily closed. The DD/DA-ratio is the ratio of the diameter of the defect and the diameter of the abdomen measured parallel to each other at the level of the defect (Fig. 1.2). The DD/DA-ratio was measured to represent the relative size of the defect in relation to the abdominal size, hypothesizing that a large defect in relation to a narrow abdomen would be difficult to close primarily. In both cases, higher ratios represent a larger defect relative to fetal size. The ratios were all measured by one investigator (T.E. Cohen-Overbeek).

We calculated the sensitivity and specificity for 5 different cut-offs per ratio and used a receiver-operating characteristics (ROC) approach to establish which cut-offs would be most suitable if the ratio was used as predictor for type of closure. We assumed the best cut-off would be the one yielding the lowest sum of false positives (FP) and false negatives (FN), i.e. inaccurate predictions, giving equivalent weight to both inaccuracies. We calculated the area under the curve as a measure of predictive accuracy, where 0.5 reflects chance prediction and 1.0 reflects perfect prediction. All calculations and statistical analyses were performed using Microsoft Office Excel 2003 and SPSS statistics version 17.0. In SPSS the ROC is approximated following the non-parametric, trapezoidal rule.

Results

Of the initial 146 cases diagnosed with an omphalocele in the selected period, 64 cases were prenatally isolated cases with an omphalocele diagnosed prior to 24 weeks of gestation. Thirty-seven cases were excluded because of TOP ($n = 22$), IUFD ($n = 12$) or a neonatal death (NND, $n = 3$). Three cases were excluded as the required image of the axial plane of the abdomen was unavailable, due to incomplete patient records and unrelated to the severity of the defect. Twenty-four cases remained (Fig. 2).

Table 1 displays the retrieved prenatal and postnatal data. Primary closure was performed in 15 of 24 cases. In 6 out of these 15 infants associated anomalies were detected after birth. Four

Table 1
Patient characteristics.

GA	Prenatal associated anomalies	Liver in omphalocele prenatally	OC/AC-ratio	DD/DA-ratio	Delivery mode, GA (weeks)	Birth weight (grams)	Type of surgery	Postnatal associated anomalies	Hospital stay (days)
19	–	No	0.53 (61/116)	0.6 (21/35)	VD, 39.4	4100	Primary	BWS	5
19	TS	No	0.53 (83/158)	0.37 (17/46)	VD, 35.0	1820	Primary	CoAo	57
19	SUA	No	0.57 (72/125)	0.53 (20/38)	VD, 38.5	2970	Primary	–	15
20	–	No	0.56 (103/184)	0.42 (28/66)	CS, 41.1	2830	Primary	–	7
20	–	No	0.39 (52/135)	0.41 (17/41)	CS, 35.5	2485	Primary	–	7
20	–	No	0.29 (34/116)	0.26 (10/38)	VD, 35.6	1919	Primary	–	15
20	–	No	0.48 (71/148)	0.25 (12/48)	CS, 33.5	2565	Primary	BWS	62
21	–	No	0.49 (82/166)	0.32 (17/54)	VD, 37.5	3650	Primary	BWS	9
22	–	No	0.72 (80/111)	0.23 (8/35)	VD, 37.1	3890	Primary	BWS	20
23	–	No	0.29 (51/176)	0.40 (12/52)	CS, 39.1	3195	Primary	–	5
23	–	No	0.4 (66/164)	0.29 (15/52)	VD, 39.0	3500	Primary	–	5
19	(twin)	Yes	0.67 (78/118)	0.53 (17/32)	VD, 37.1	2200	Primary	–	9
20	–	Yes	0.76 (97/128)	0.48 (20/42)	VD, 33.6	1960	Primary	Small VSD	18
20	–	Yes	0.81 (122/150)	**0.7 (35/50)***	VD, 34.2	3430	Primary	–	20
21	–	Yes	0.59 (86/145)	0.46 (21/46)	VD, 38.6	3100	Primary	–	25
16	–	Yes	1 (60/60)	**0.52 (12/23)****	VD, 38.6	2730	Delayed	ACC, AVSD	100
19	–	Yes	0.94 (117/124)	0.64 (23/36)	VD, 35.6	2600	Delayed	–	417
20	–	Yes	1.02 (134/132)	**0.50 (21/42)****	VD, 30.6	1495	Delayed	–	106[a]
21	–	Yes	0.90 (128/142)	0.65 (30/46)	VD, 35.4	1870	Delayed	ToF	57[a]
21	–	Yes	0.95 (115/121)	0.73 (27/37)	CS, 38.5	3000	Delayed	–	49
22	–	Yes	**0.74 (115/155)****	0.67 (30/45)	CS, 39.3	2140	Delayed	Mild AoVS	108
22	–	Yes	0.91 (106/116)	0.86 (32/37)	CS, 30.0	1500	Delayed	–	192[b]
22	–	Yes	0.83 (135/163)	0.69 (36/52)	CS, 39.1	5390	Delayed	BWS, mVSD	42
23	–	Yes	0.93 (136/147)	**0.5 (22/44)****	VD, 38.6	2910	Delayed	–	23

Legend: ACC, Agenesis of the corpus callosum; AoVS, Aortic valve stenosis; AVSD, atrial ventricular septal defect; BWS, Beckwith-Wiedemann Syndrome; CoAo, Coarctation of the Aorta; CS, caesarean section; DD/DA, ratio of the defect diameter and the abdominal diameter; GA, gestational age in weeks. days; mVSD, musculous ventricular septal defect; OC/AC, the ratio of the omphalocele circumference and the abdominal circumference; SUA, Single umbilical artery; TS, Turner Syndrome; ToF, Tetralogy of Fallot; VD, vaginal delivery; VSD, ventricular septal defect.

[a] Infant death at 4 months.

[b] Infant death at 6 months.

* False positive (FP).

** False negative (FN) for type of closure after calculating the cut-offs.