


Fetal cephaloceles: prenatal diagnosis and course of pregnancy in 65 consecutive cases

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Abstract

Purpose To scrutinize the characteristics of all cases with antenatally established diagnosis of cephalocele in two tertiary referral centers.

Methods We retrospectively reviewed 65 cases diagnosed with cephaloceles and tabulated sonographic findings and autopsy recordings as well as medical charts of all survivors in terms of clinical outcome.

Results The case notes of 65 fetuses were available for final analysis. Gestational age (GA) at diagnosis ranged from 10.4 to 38.1 weeks. Of our cohort, 53/65 cases (80%) had occipital protrusions, 10 (15%) were found to have frontal lesions, and another two had parietal cephaloceles. A total of 52 pregnancies were terminated or resulted in intrauterine fetal demise (78%). In 18 cases (11%), the cephalocele was part of underlying syndromic disorders (e.g., Meckel–Gruber syndrome). Thirteen pregnancies were continued until term, out of which all affected

individuals were live-born. Neurosurgical intervention was prompted within the first 7 months postnatally.

Conclusions In general, the outcome of fetuses with cephaloceles is rather poor as four out of five pregnancies were terminated. Postnatal outcome of all survivors in our cohort was rather determined by localization of the cele and more important by the presence and severity of concomitant malformations than the extent of the lesion.

Keywords Encephalocele · Meningocele · Cephalocele · Prenatal · Ultrasound · Outcome

Introduction

Cephaloceles—cranial meningoceles and encephaloceles—are characterized by a skull defect enabling herniation of intracranial tissue [1]. Their incidence is estimated 1:2000 to 1:5000 live births with marked geographic and ethnic differences. Whereas the prevalence of neural tube defects has substantially decreased since folic acid fortification has been recommended in the early 1990s, this issue has been controversially debated in terms of the development of cephaloceles [2, 3]. The underlying etiology of cranial herniations is not fully understood, but includes ethnic, genetic, and environmental factors. They merely constitute a failure of separation of the neural and surface ectoderm after the closure of the rostral neuropore has been completed during the 4th gestational week [4]. As previous studies suggested, the prognosis strongly depends on the anatomical site of the lesion, the presence and extent of neural contents as well as coexisting malformations [3]. It ranges from unremarkable physical and cognitive development to severe neuromotoric disabilities. It has been further shown that anatomic disruption and cognitive and

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physical impairment do not necessarily correlate [5]. Therefore, precise prenatal diagnosis as early as possible is indispensable in order to provide the expectant parents with adequate advice. The detection rate of cephaloceles during first trimester has been estimated 80%, they constitute 5% of all detected severe structural defects of the central nervous system (CNS) [1, 6]. Associated malformations or conditions comprise hydrocephalus, which is associated with the worst prognosis, Meckel–Gruber syndrome, trisomy 18, single-gene disorders, Walker–Warburg syndrome, cerebro-ocular-muscular syndrome (COMS), and toxic embryopathies [3].

The aim of our work was to evaluate the incidence, pre- and postnatal course and additional anomalies of prenatally detected cephaloceles in a mixed low- and high-risk population. The present retrospective study illustrates the issue of prenatal diagnosis of cephalocele and aims at correlating prenatal diagnostic findings with postnatal development or autopsy outcome of affected fetuses registered in two tertiary referral centers for prenatal medicine.

Methods

This retrospective study included all pregnancies with an abnormal appearance of the fetal CNS diagnosed antenatally by ultrasound examination between the years 1994 and 2012. Screening for fetal CNS abnormalities as an integral part of fetal anomaly survey is routinely offered to all pregnant women attending our prenatal referral centers at the University Hospital of Schleswig-Holstein, Campus Lübeck, and at the University Hospital Bonn. In our mixed-risk collective, the indications for ultrasound examination varied from first and second trimester anomaly screening to referral due to family history and due to suspected CNS anomalies for further investigation. All ultrasound examinations were performed with an Acuson Sequoia 128 XP (Siemens, Erlangen, Germany), an ATL HDI 5000 (Philips, Solingen, Germany), a Voluson 730 Expert (GE Healthcare, Milwaukee, USA), or IU22 (Philips, Solingen, Germany) ultrasound machines using a 2–5, a 4–7, or

5–9 MHz curved array abdominal transducer or a 4–8 MHz vaginal sector probe.

During the study period, all pregnancies with cephaloceles (encephaloceles and cranial meningoceles) were tabulated for further analysis. 65 pregnancies diagnosed between the years 1994–2012 were reviewed including prenatal imaging, postnatal studies, operative findings, pediatric charts of the survivors, and autopsy reports for terminated pregnancies. The average postnatal follow-up was 12 months. Necropsies after termination of the pregnancy (TOP), stillborn, or neonatal death were performed by experienced pathologists trained in fetal autopsy.

Maternal characteristics are summarized in Table 1. Emphasis must be placed on the history of precedent miscarriages, which was slightly increased (37%) in comparison to the German population (approximately 30%).

Results

During the study period, a total of 65 pregnancies were diagnosed with fetal cranial tissue herniation. Mean gestational age at diagnosis was 18 + 0 weeks of gestation (ranging from 10.4 to 38.1 weeks). There was a slight female preponderance (34 fetuses vs. 30 males). In one fetus, the gender could not be determined. Karyotyping was conducted in 51 of 65 cases. Chromosomal anomalies were found in three cases with, trisomy 18 (47, XX, +18), triple X syndrome (47, XXX), and a chromosomal deletion (46, XX, del(14)(q24-qter)), respectively. The remainder (45 of 51 cases) had normal chromosomes. Single-gene disorders were present or suspected in 18 fetuses: Meckel–Gruber syndrome (MKS) was diagnosed in 11 cases (affected siblings in three cases). In three fetuses of the same family, occipital encephalocele was diagnosed as a part of oro-facial-digital syndrome (OFDS) type IV (out of which two cases were already published by Rösing and co-workers [7]). In one family, two fetuses showed small occipital meningocele as a part of Joubert-related disorder, in one family with a previous affected sibling the fetus showed a small occipital encephalocele as a part of

Table 1 Maternal and birth characteristics

Maternal and birth characteristics	Range [number]	Median [percentage]
Maternal age (years)	[18–40]	30.3
Maternal body mass index (kg/m ²)	[15.8–36.4]	26.1
Parity (nulliparous)	[28/65]	[43%]
History of abortion	[24/65]	[37%]
History of precedent malformations	[11/65]	17%
Gestational age at first diagnosis (weeks)	[10.4–38.1]	18.0
Number of ultrasound examinations	[1–7]	2.7
Live-born infants	[13/65]	20%

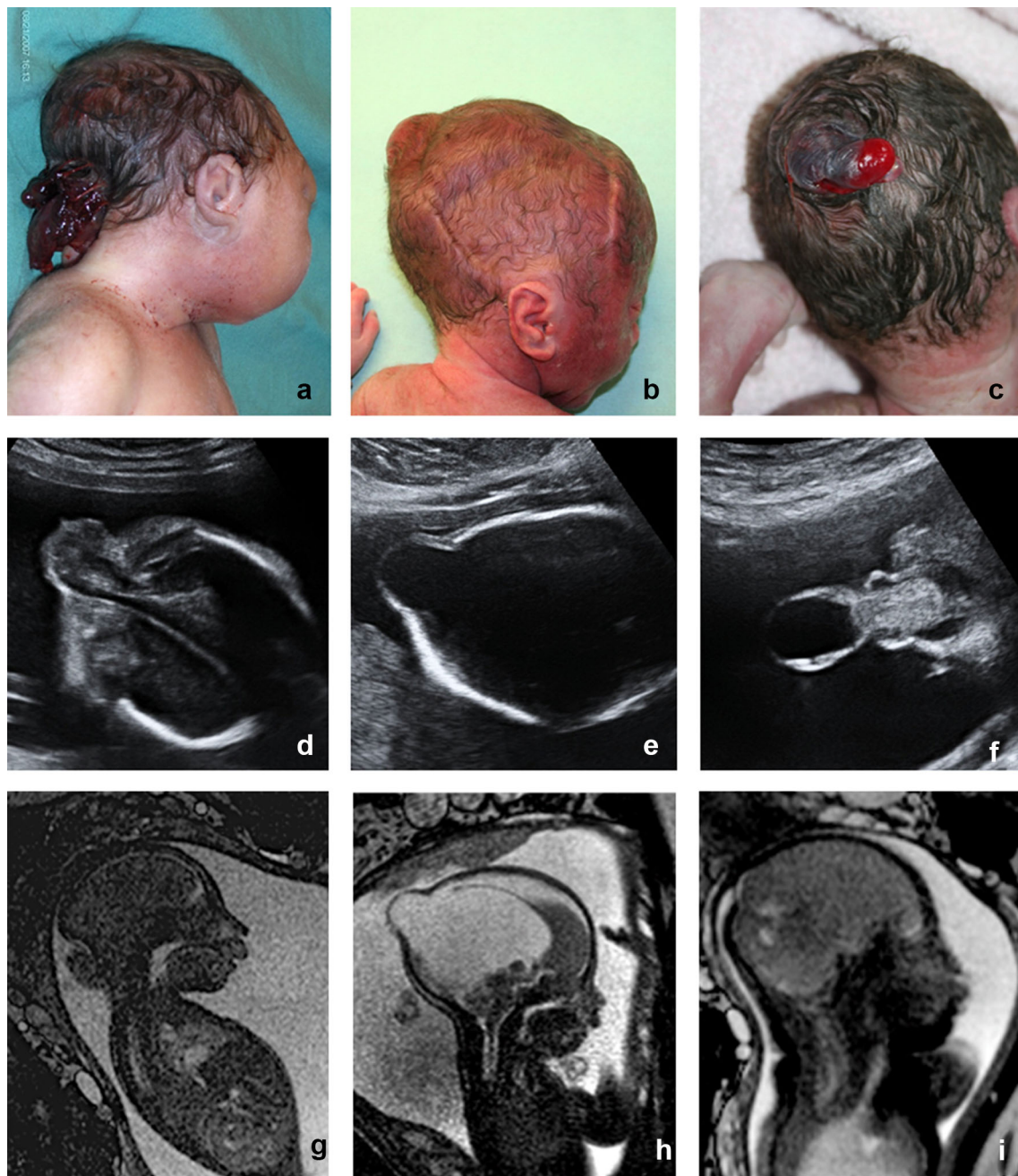


Fig. 1 Different appearance of fetal cephaloceles. Occipital encephalocele at 30 weeks with marked posteriorly located cranial protrusion (a), corresponding transversal sonographic view (d) and MR image (g) both depicting a bony lesion enabling herniation of intracranial (cerebellar) tissue. The fetus was stillborn at 33 gestational weeks, the parents declined autopsy because of religious

objections. Images b, e, h showing an occipital meningocele at 28 weeks clearly lacking any neural content. Parietal encephalocele at 33 weeks (c) communicating with meningeal sac through a skull defect as seen both on US (f) and MR images (i). The pregnancy in the latter two cases was terminated on parental request. Pathological examination during autopsy confirmed the prenatal diagnosis

Walker–Warburg syndrome. Furthermore, one fetus affected by an occipital encephalocele showed signs of oculo-auriculo-vertebral dysplasia (Goldenhar-Gorlin syndrome). A single umbilical artery was found in 4 of the 65 study cases.

Fifty-three (80%) of the 65 fetuses had cephaloceles located occipitally, from those three fetuses had

craniospinal dysraphism as part of disruptive anomalies (amniotic band sequence). Ten (15%) showed frontal lesions (eight with nasofrontal protrusion, one with infraorbital protrusion, and one with a nasoethmoidal protrusion), and two (3%) had a parietal lesion (Figs. 1, 2). Termination of pregnancy on parental request was undertaken. In 47 of 65 cases (72%) herniated brain tissue was

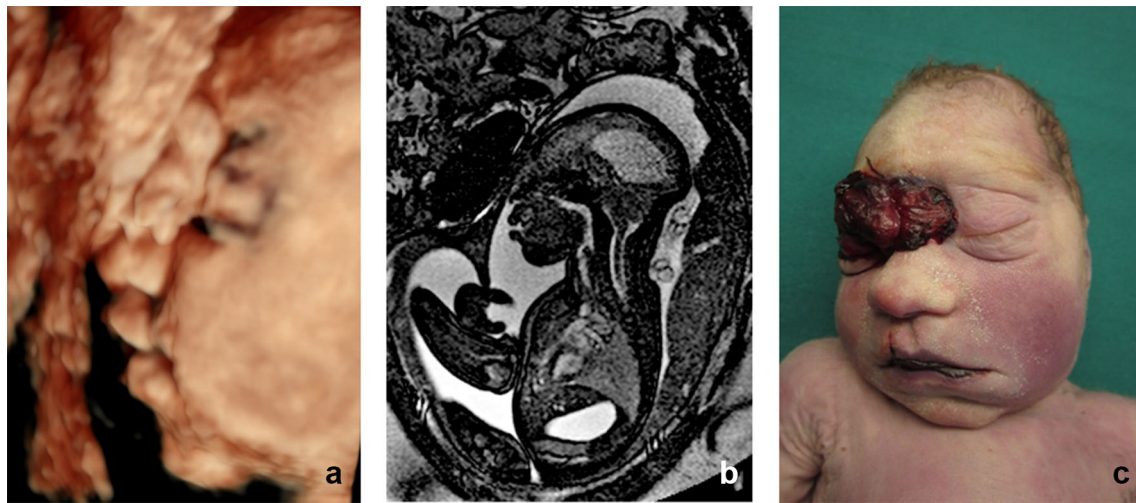


Fig. 2 Frontoethmoidal encephalocele at 34 gestational weeks. Rendered 3D image showing frontal protrusion (a) and corresponding MRI with herniated intracranial tissue (b). Gross appearance on autopsy, note the marked microcephalus (c)

visualized. Termination of the pregnancy on parental request was undertaken in 49 of 65 cases (75%) (among those, selective feticide was performed in two dichorionic twin pregnancies in which the unaffected co-twins were born by Cesarean section with favorable outcome), and intrauterine fetal death occurred in 3 of 65 (5%) cases. All couples were extensively counseled (together with neuro-pediatricians and neurosurgeons). The presence of accompanied cerebral and associated extracerebral anomalies was the main cause for parents to opt for termination of the pregnancy.

In 13 of 65 (20%) pregnancies with prenatally detected cephalocele, in whom the parents opted for continuation of the pregnancy, prenatal short-term follow-up examinations were scheduled, and in 11 cases Cesarean section was planned at 38 completed weeks (Table 2). In another two cases, the mode of delivery was vaginal birth. All 13 children were live-born without gross impairment during neonatal period (all newborns presenting with 10-min Apgar scores of 9 or 10, respectively). One of the survivors had an additional X chromosome as stated above. In all cases, MRI was performed postnatally (prenatal MR imaging was available in four of these fetuses) and subsequent surgical interventions were conducted. Postnatal neurosonography and MRI confirmed prenatal diagnosis in all live-born fetuses. Neurosurgery was performed within the first 7 months postnatally. Outcome was strongly depending on the severity of concomitant cerebral anomalies.

Long-term pediatric follow-up was available in two of the survivors. In one case, postnatal and long-term outcome was rather favorable in spite of severe cerebro-cranial malformations that had been diagnosed antenatally. The child suffered from cranio-cervical meningocele, hypoplasia of the cerebellum, cervicothoracic syrinx, and

complex cervico-spinal malformation. At the age of two weeks, neurosurgery was performed. At the age of 3 9/12 years, the child showed diminished cognitive and motor skills (particularly speech development). However, skills improved significantly with ergotherapy and logopedic support. Neurologic status was uneventful apart from discrete dysdiadochokinesis. The second case with frontoethmoidal encephalocele and additional arachnoidal cyst had neurosurgical resection of the cele at the age of 7 weeks. The concomitant arachnoidal cyst was resected at the age of 7 months. The girl showed impaired motor and cognitive skills and presented with seizures and macrocephaly and plagiocephalus. At the age of 18 months, the child demonstrated slightly retarded motor development and normal neurologic state.

In the remaining 11 cases, postnatal follow-up was available up to the age of 6–12 months. In all, but one case (tiny parietal meningocele) surgical resection of the herniated tissue and plastic covering of the defect was necessary. Four children needed additional ventriculoperitoneal shunting due to concomitant hydrocephalus. So far, all cases solely affected by meningeal protrusions showed a normal or only slightly impaired development (Table 2). Those infants who had resection of an encephalocele (containing cerebral tissue) were more likely to have neuromotoric sequelae. However, based on our data, there were no severely disabled individuals to report.

Discussion

This is a large cohort study of antenatal cases diagnosed with cephaloceles. The majority of fetuses (72%) enrolled in this study suffered from herniated brain tissue. Out of

Table 2 Live-born fetuses with antenatally diagnosed cephalocele

Case	Karyotype	GA (weeks)	Location	Size (mm)	Ratio (cele/BPD)	Content	CNS anomalies	Other anomalies	Delivery	Details	Outcome
1	46, XY	22 + 3	Parietal	10 × 10	0.18	Meningocele	–	–	Vaginal birth 40 + 4	–	No surgery required; normal development
2	Female*	22 + 6	Occipital	24 × 23	0.47	Cerebral tissue	Microcephalus, Chiari II	–	C/S 32 + 0 (PROM)	–	Microcephaly, surgical coverage; until age 7 months no major impairment
3	46, XX	24 + 3	Occipital	20 × 20	0.30	Meningocele	Hydrocephalus internus	SUA	C/S 35 + 1	–	Surgery, VP shunt, normal development
4	46, XX	21 + 5	Occipital	9 × 9	0.18	Meningocele	–	–	C/S 37 + 6	sibling with renal agenesis	surgery at 6 months; normal development
5	46, XX	28 + 5	Occipital	21 × 22	0.32	Meningocele	–	–	C/S 36 + 5	MC twins	Surgery at 6 months; normal development at 12 months
6	Female*	32 + 5	Occipital	66 × 56	0.89	Meningocele	Asymmetric hydrocephalus	–	C/S 37 + 5	–	Meningocele, surgery, VP shunt, normal development
7	46, XX	20 + 6	Occipital	23 × 20	0.47	Cerebral tissue	Hydrocephalus, Chiari II microcephalus	–	C/S 37 + 1	–	Microcephaly, surgery, mild delay at age 12 months
8	47, XXX	22 + 2	Occipital	21 × 21	0.40	Cerebral tissue	Microcephalus, hydrocephalus internus	Perimembranous VSD*	C/S 37 + 1	–	Surgery, VP shunt; postnatal confirmation ACC, VSD, ASD II*, mild developmental delay
9	Female*	23 + 6	Occipital	19 × 17	0.35	Cerebral tissue	–	–	C/S 37 + 5	–	Surgery (cerebral tissue and meninges)
10	46, XY	23 + 4	Occipital	23 × 28	0.46	Meningocele	–	–	CD 38 + 2	–	Surgery; mild delay of neuromotoric development at age of 4 years
11	Male*	32 + 5 (infraorbital)	Frontal	40 × 42	0.44	Cerebral tissue	–	–	C/S 37 + 2	–	Favorable outcome after surgery, good motoric, social and cognitive development
12	46, XY	21 + 1	Occipital	25 × 33	0.66	Meningocele	Chiari II	VSD	C/S 38 + 0	Vitamin A intake	Surgery at age 6 months, VP shunt, hypoplastic cerebellum, mild delay
13	46, XX	34 + 1	Frontal	26 × 18	0.31	Cerebral tissue	Arachnoidal cyst	–	Vaginal birth 38 + 4	–	Surgery, arachnoidal cyst, moderate motoric retardation at age 8 months

ASD atrial septal defect, ACC agenesis of corpus callosum, BPD biparietal diameter, C/S Cesarean section, CNS central nervous system, MC monochochonic, PROM premature rupture of membranes, SUA singular umbilical artery, VP shunt ventriculoperitoneal shunt, VSD ventricular septal defect

* Phenotype, no antenatal karyotyping performed

which, only six pregnancies continued until term, whereas nearly half of fetuses who had solely meningeal protrusions were live-born. Short-term (and in two cases long-term) follow-up revealed that sixty percent of the survivors showed normal development or only had a mild neuro-motoric delay.

Prenatal sonographic diagnosis in most cases is based on the direct visualization of the cephalocele and the osseous defect, which is typically localized in the midline and rarely lateral in association with the cranial sutures. Overall, occipital cephaloceles constitute 75% of all craniocerebral bulgings [3]. In our patient population, we noticed a similar distribution (80% occipital defects vs. 6% parietal lesions). Another 14% were frontally located, which is in line with previous reports (Table 3). Interestingly, the latter are discussed to constitute not a genuine form of neural tube defect (NTD), but rather originate from environmental influences (e.g., reduced folic acid intake) and are more common in Southeast Asia [2]. In this regard, maternal passive smoking has recently been attributed to be an independent risk factor for three subtypes of NTD (anencephaly, spina bifida and most strongly encephalocele) [17, 18]. An occipitocervical lesion accompanied by features of Chiari II malformation (herniation of posterior fossa contents) has also been described as Chiari type III malformation (present in two live-born fetuses in our study) [19, 20].

Although the onset of cephaloceles reportedly occurs between 25 and 50 days of gestation (for anteriorly located lesions) and up to 60 days (for posterior defects), respectively, the median GA in our study at initial diagnosis was 18 + 0 gestational weeks (Table 2) which is in accordance to current literature [14]. In 43% the diagnosis was established before 20 completed weeks. The earliest GA at diagnosis was 10 + 2 weeks (together with 14 additional cases identified during first trimester). Recently, Sepulveda and co-workers stated that with detailed evaluation of skull contour all cases of cephaloceles are potentially amenable to early first trimester diagnosis [15]. An early diagnostic hint might be an enlargement of the rhombencephalic cavity, a structure that is readily seen between 8 and 12 gestational weeks [21].

Although cephaloceles as small as 3 mm in diameter could be detected during first trimester scan, the issue of correct antenatal diagnosis of any neural involvement remains challenging. Moreover, one has to bear in mind that progression from meningocele to meningoencephalocele with advancing pregnancy has been described [6, 22]. On the other hand, regression of an encephalocele has also been found [23]. Bronshtein and Zimmer reported on a case with a changing appearance of the cele (sliding of herniated brain tissue in a to and fro manner) [24]. Nevertheless, this has to be taken into account during counseling of the parents after early diagnosis of a cephalocele. It is of note

that in our series, similar to Sepulveda's findings (two live-borns), none of the affected pregnancies continued until term after early diagnosis of a cephalocele. Although there was a certain increase in relative size throughout pregnancy, we did not notice any change in gross appearance during repeated scans.

It has recently been shown that in the case of open spina bifida, as a more frequent neural tube defect, a biparietal diameter <5th percentile will tenfold increase the likelihood of a spinal dysraphism [25]. Applying these results on our study population, microcephaly was not a consistent finding, but if present it did worsen the prognosis as stated beneath. The main differential diagnoses of cephaloceles include cystic hygroma, teratoma, hemangioma, and scalp edema [26–29]. In this regard, detection rates of cephaloceles are frequently higher in case of high-risk family constellation, (e.g., affected relatives with Meckel–Gruber, Walker–Warburg, oro-facial-digital syndrome, Joubert syndrome and related disorders (JSRD), or Knobloch syndrome, respectively, as in the present study) [30]. In fact, in 13 of 65 cases enrolled in our study, a genetic predisposition was registered (consanguinity, affected siblings or family). However, familial recurrence of isolated non-syndromic cephalocele is an uncommon phenomenon [31] and was present in a single family in our series. Definitive prenatal diagnosis might be achieved by applying molecular diagnostic techniques (e.g., targeted DNA sequencing chromosomal microarray, and whole genome sequencing) [32, 33].

In isolated cephaloceles, the volume of protruding brain parenchyma and consecutive microcephaly constitutes the main impact on prognosis postnatally. There were several attempts for categorizing craniocerebral protrusions in either prenatal and postnatal cohorts, all of which tabulated affected individuals with respect to their gross appearance (cystic vs. solid nature or classification based on the origin and/or extent of herniated tissue) [5, 34–36]. Bannister et al. pointed out that large herniation sacs were not necessarily associated with a large mass of herniated tissue, and thus the size of the entire cele does not correlate with the prognosis for the fetus [14]. These findings are closely related to our observations, as we were able to show that the size of the cele (ranging from 5 × 6 to 66 × 56 mm) did not allow to draw reliable conclusions regarding both the course of pregnancy and the postnatal outcome in our cohort. Although 12/13 live-born fetuses needed surgical treatment, even those two children with larger herniations had no signs of severe developmental delay. On the other hand, it has been proposed that if >50% of intracranial tissue is herniated, postnatal survival is rather unlikely [13]. A large neurosurgical study revealed that after surgical intervention 50/85 patients (59%) showed normal development or only had mild delay, 14 (16%) had moderate, and 21 (25%) had severe delay, respectively [35].

Table 3 Overview of available studies assessing prenatal cases of cephaloceles

Study	Population	Country	period	GA (weeks)	n	f ^a	Par	Occ	Details	Outcome
Graham et al. [8]	Prenatal	USA	n/a	22–24	5	1	–	4	Hydrocephaly (1)	TOP (3), ND (1), alive (1)
Simpson et al. [9]	Prenatal	AU	1975–1983	22	4	–	1	3	DWM (1)	TOP (2), alive (2)—1 disabled, 1 well/shunt
Chervenak et al. [10]	Prenatal	USA	1981–1982	19–35	8 ^a	–	1	6	–	TOP (5), ND (1), alive (2; disabled/shunt)
Goldstein et al. [4]	Prenatal	USA	1984–1989	15–40	15	1	1	13	VM (7), ACC (2), cleft (2), MCKD (2), aneuploidy (4)	TOP (5), IUFD (2), ND (4), alive (3; disabled)
Jeanty et al. [11]	Prenatal	USA	1986–1988	15–35	15	2	2	11	MCKD (1), cleft (1), Klippel–Feil syndrome (1)	TOP (9), ND (2), alive (4; 2 disabled, 2 well/surgery)
Winninger et al. [12]	Prenatal	USA	1985–1993	12–40	15	1	1	13	Aneuploidy (2), PC (1), cleft (2), WWS (1), MKS (1), DWM (1)	TOP (6), IUFD (1), ND (3), alive (5; 3 disabled after resection, 2 lost)
Budorick et al. [13]	Prenatal	USA	1978–1991	13–36	24	4	3	17	VM (6), aneuploidy (1), MKS (5), DWM (2), ABS (3), cleft (1)	TOP (17), ND (4), death <1 year (2), alive (1 well/surgery)
Bannister et al. [14]	Prenatal	UK	1991–1997	18–20	31	4	1	26	VM (4), DWM (4), hydrocephaly (1), MKS (8), cleft (3)	TOP (19), IUFD (4), ND (1), alive (7; 6 well, 1 disabled)
Sepulveda et al. [15]	Prenatal	CHL	2002–2014	11–14	35 ^b	3	3	27	MKS (4), ABS (2), skeletal dysplasia (1)	TOP (21), IUFD (4), ND (1), lost (8), alive (1 well/surgery)
Kasprian et al. [16]	Prenatal	A	2004–2013	18–30	14	–	–	14	WWS (1), Joubert syndrome (1)	ND (2), IUFD (1), lost (5), alive (6; 5 disabled, 1 well)
Present study	Prenatal	D	1994–2012	10–38	65	9	4	52	MKS (11), DWM (5), aneuploidy (3), ABS (3), Joubert-like syndrome. (1), OFDS (3), ACC (3), VM (2), PC (1), hydrocephaly (19), Goldenhar–Gorlin-like syndrome (1)	TOP (49), IUFD (3), alive (14);
Total	Prenatal	–	1975–2015	10–40	234	25	17	187	MKS (29), hydrocephaly (21), VM (19), DWM (13), aneuploidy (10), ABS (9), cleft (7), ACC (5), OFDS (3), MCKD (3), WWS (2), PC (2), Joubert-like syndrome (2), Goldenhar–Gorlin-like syndrome (1), Klippel–Feil syndrome (1), skeletal dysplasia (1)	TOP (136), ND (19), IUFD (12), death <1 year (2), alive (46), lost (14)

ABS amniotic band sequence, ASD atrial septal defect, ACC agenesis of corpus callosum, DWM Dandy–Walker malformation, GA gestational age, GDM gestational diabetes mellitus, IUFD intrauterine fetal death, MCKD multicystic kidney disease, MKS Meckel–Gruber syndrome, ND neonatal death, not determined, OFDS orofaciocaudal syndrome, PC pentalogy of Cantrell, TOP termination of pregnancy, VM ventriculomegaly, WWS Walker–Warburg syndrome

^a In one case cephalocele protruded through a grossly malformed skull

^b In two other cases cephalocele was part of a disruptive lesion and involved several areas of the skull

Focusing on the impact of location of the skull defect, occipital encephaloceles are supposed to be associated with a higher frequency of hydrocephalus and seizures during postnatal life [3].

Although fetal neurosonography remains the primary imaging modality, magnetic resonance imaging (MRI) as an adjunctive tool to detailed CNS examination might be of help particularly in those cases with limited acoustic window. According to Kasprian et al., MR imaging is of value in assessment of gyration anomalies and brainstem morphology of externalized brain tissue as this has been reported to be subject of degenerative changes throughout pregnancy [16]. In our patient population, fetal MRI was performed beyond 20 completed weeks of gestation in eight fetuses out of which four were live-born (Fig. 1). There was no additional information added by MRI in these fetuses compared to the results of detailed neurosonography. Nevertheless, in case of continuation of the pregnancy or when additional cerebral malformations are suspected by ultrasound, fetal MRI is an adjunct for establishing a precise diagnosis.

Our study has some limitations which have to be pointed out. First the retrospective design of the study; second there is a certain bias because of the referral population focusing on more severe cases; third there is only a small number of pregnancies continued until term. Furthermore, the follow-up is limited and no systematic approach for assessment of the surviving infants was available.

Conclusions

In case of prenatally suspected cephalic anomalies (increased nuchal translucency, small skull (reduced biparietal diameter), enlarged rhombencephalic cavity), affected fetuses in prior pregnancies or a positive family history, early diagnosis of cranial herniations as early as the first trimester is feasible. After establishing the diagnosis, a thorough evaluation of accompanying anomalies of the fetus is mandatory and invasive testing should be offered including advanced molecular genetic analysis. Although most of the affected pregnancies were terminated (Table 3), 20% the parents opted for continuation of the pregnancy. In our cohort, there were no severe neuromotoric disabilities (after neurosurgical intervention) reported. Parental counseling should be individually tailored in an interdisciplinary approach together with neuropediatricians, geneticists, and neurosurgeons. In those cases, repeated sonograms and MRI are strongly recommended.

Compliance with ethical standards

Funding This study was not funded.

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors. It is a retrospective archive study. The institutional review boards of both the universities of Bonn and Lübeck do not require formal approval for retrospective archive studies; therefore, an ethical approval was not sought.

Informed consent Informed consent was obtained from all individual participants enrolled in this study.

Author contributions UG, JW: Protocol/project development. UG, MK, UG, RAF, AK, AG, ChB, JW: Data collection or management. JW, FH, UG: Data analysis. JW, FH, UG: Manuscript writing/editing.

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