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Anencephaly and obstetric outcome beyond the age of viability

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Abstract

Objective: To review the obstetric impact and natural history of anencephalic pregnancies beyond the age of viability.
Study design: A retrospective chart review of all cases with a prenatal diagnosis of anencephaly who delivered after 24 weeks' gestation during the period 1990 until 2016. Obstetric outcomes including mode of delivery, live births, shoulder dystocia, antepartum haemorrhage (APH), postpartum haemorrhage (PPH) and uterine rupture were studied.
Results: A total of 42 cases were studied. The average gestational age at diagnosis was 22 weeks (range 10–41). The average gestational age at birth was 36 weeks (range 25–44 weeks). Induction of labour was performed in 55% (23/42) of the cases. Livebirths were documented in 40% (17/42) of the cases. The average birth weight was 1597 ± 746 g. The rate of vaginal birth was 69% (29/42), the overall rate of caesarean section was 31% (13/42), with a primary caesarean section in 31% (4/13) and a repeat caesarean section in 69% (9/13) of the patients. There were two cases of shoulder dystocia. No other complications were encountered.
Conclusion: Overall, anencephaly is not associated with an increased risk of obstetric complications; however, there is a tendency towards delivery via repeated caesarean section in women with a previous uterine scar and anencephaly. The prenatal counselling of potential obstetric outcomes could be of robust value for parents who opt to continue with anencephalic pregnancies.

Keywords: Anencephaly; antepartum haemorrhage (APH); caesarean section; early fetal anatomy; induction of labour; postpartum haemorrhage (PPH); prenatal counselling; prenatal diagnosis; shoulder dystocia.

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Introduction

Anencephaly is a severe defect of the development of the neuroaxis, in which the developing forebrain and variable amounts of the brainstem are exposed *in utero*, due to the failure of the neural tube to close at the base of the skull in the third or fourth week (days 26–28) after conception [1–3]. It is an invariably lethal condition with no possible intervention or treatment; although some fetuses are born alive, most of them die within a few hours after birth [4]. There have also been reports of prolonged period of life; some anencephalic infants were considered as prospective organ donors and therefore survived for a week after being provided with intensive care support [5, 6]. In most cases of death in anencephalic infants, cardiorespiratory arrest apparently occurs before the cessation of brainstem functions [7].

According to data from birth certificates reported to the National Center for Health Statistics, the prevalence of anencephaly in the US in 2001 was 9.40 per 100,000 live births [8]. The incidence tends to appear higher in countries where abortion is legally restricted [9].

Locally the cause of having such a severe anomaly beyond the age of viability could be related to parents' religious beliefs and restricted legal termination rules in addition to other factors such as late ultrasound detection and delay in performing a fetal anatomy scan. We aimed in our study to review the obstetric impact and natural history of anencephaly beyond the age of viability, and thus to optimise the prenatal counselling.

Materials and methods

We performed a retrospective cohort study of women who delivered singleton infants with isolated anencephaly after 24 weeks' gestation at our institution during the period 1990–2016 inclusively. The diagnosis of anencephaly was made by a prenatal ultrasound and confirmed by a postnatal neonatologist examination.

Data were collected from the patients' medical records, ultrasound reports and our fetal medicine database.

Outcome variables included gestational age at diagnosis, gestational age at birth, maternal diabetes, induction of labour, caesarean delivery, live births, stillbirths, shoulder dystocia, uterine rupture, antepartum haemorrhage (APH) and postpartum haemorrhage (PPH).

The study was approved by the hospital's Institutional Review Board.

Results

A total of 42 patients who delivered anencephalic fetuses after the age of viability during the study period were identified and studied. The average maternal age at birth was 28 years (range 20–42). The average parity was 2 (range 0–9). Twenty-one percent (9/42) of the patients were primigravidae. Maternal pre-existing diabetes was observed in 12% (5/42) of the patients. The gestational age at diagnosis was 22 weeks (range 10–41 weeks). Seventeen percent of these cases (7/42) were detected prior to 18 weeks' gestation, while the gestational age at birth was 36 weeks (range 25–44 weeks).

The average birth weight [mean ± standard deviation (SD)] was 1597 ± 746 g (range 530–3190 g). Stillbirths were reported in 60% (25/42) while the fetuses were born alive in 40% (17/42) of the patients; however, all died within the first day of life.

Vaginal birth was achieved in 69% (29/42) of the patients. Induction of labour was performed in 55% (23/42) of the patients. There were no differences in achieving vaginal birth in the group who had induction of labour compared to spontaneous labour with a rate of caesarean section of 13% (3/23) compared to 10% (1/10), respectively (P-value = 1).

The overall rate of caesarean section in our cohort was 31% (13/42), with a primary caesarean section in 31% (4/13) and a repeat caesarean section in 69% (9/13) of the patients. The indications for primary caesarean sections were failed induction in two cases, abnormal lie in one case and shoulder dystocia in one case, in which after a failed trial of delivering the foetal body using standard manoeuvres for shoulder dystocia management, which included McRoberts manoeuvre, suprapubic pressure, internal rotational manoeuvres and delivery of the posterior arm, the fetal body was delivered through cephalic replacement into the pelvis followed by a caesarean section (Zavanelli manoeuvre). There were a total of two cases of shoulder dystocia of which none were diabetic. There were no reported cases of APH, PPH, complicated caesarean sections and uterine rupture.

Discussion

Based on our local rules and ethical regulations, termination of pregnancy is not entirely prohibited. In fact, termination is permissible after being reviewed by the hospital ethics board in cases of a confirmed lethal or severe fetal anomaly but the gestational age is limited to 19 weeks or less. Therefore, some women would have missed the chance of being offered termination of pregnancy if the diagnosis was made beyond that gestational age; in addition, some parents might opt to continue with pregnancy regardless of the pregnancy outcome for various social and/or religious reasons.

It is important to address with the parents all the potential possibilities of maternal outcome especially when the termination of pregnancy is not or no more an option.

In our cohort, the average gestational age for ultrasound detection of anencephaly was 22 weeks which is around the recommended gestational age for the fetal anatomy survey.

Prenatal detection of anencephaly by ultrasound is high and reliable at 10–14 weeks' scan [10, 11]; however, less than one fourth of our anencephaly cases were detected by ultrasound prior to 18 weeks' gestation. Lack of adherence to a universal structured foetal screening programme within the government and private sector and perhaps lack of public awareness might be potential causes of a late maternal presentation. The International Society of Ultrasound in Obstetrics and Gynaecology (ISUOG) in 2011 recommended that countries where pregnancy termination is restricted should balance detection rates against the time needed for counselling and investigation [12].

Despite the routine anatomy screening being at 20 weeks' gestation, Blaas [13] reported that some major anomalies, such as anencephaly, can be diagnosed as early as 9 weeks. However, the diagnosis at this very early gestational age is still unreliable. Nevertheless, the first trimester screening at 11–14 weeks would provide more reliable information about the foetal anatomy, which is comparable in several aspects to anatomy scans performed in the second trimester. The sensitivity of first trimester anatomy screening significantly increases with gestational age (from about 45% at 11 weeks to 75% at 14 weeks) and strong differences exist between the organ systems [14–16].

An early anatomy ultrasound will detect structural nongenetic defects and defects associated with genetic anomalies other than the common trisomies [17].

The additional value of an early foetal anatomy allows expecting parents to provide informed decision regarding further management of their pregnancy, that includes the option of pregnancy termination [18]. It also allows them more time to consider their options, rather than being pressed by the legal time constraints on pregnancy termination in the late mid-trimester [17].

Countries where termination of pregnancy is limited to certain gestational age would benefit from adopting an early foetal anatomy screening program [19]. Additionally, maternal safety can be more achievable when the termination of pregnancy is opted to be done early rather than late in gestation [20].

The rate of stillbirths in our anencephaly cohort was 40% which is similar to that observed in a previously published anencephaly series by Obeidi et al. [4] of 42%. However, in the same series they reported a higher rate of shoulder dystocia of 16% (4 out of 25) while we reported shoulder dystocia in 4% (2 out of 42). Shoulder dystocia in anencephaly is not uncommon, that is possibly attributed to a diminished head size, which does not dilate the cervix enough and delivers the head with lesser dilatation of the cervix while the shoulders, though normal unlike in macrosomia, get stuck [21]. None of the shoulder dystocia patients in our cohort were diabetic, which supports that anencephaly could be an independent risk factor for shoulder dystocia.

Induction of labour might carry an increased risk for caesarean section [22]. However, in our cohort there were no differences in achieving vaginal birth in women who were induced compared to women who spontaneously laboured. That is possibly because often the standard foetal monitoring is not implemented in severe foetal anomalies such as anencephaly and therefore caesarean sections for foetal reasons do not apply based on our beneficence-based obligation to the pregnant woman to allow her to avoid caesarean delivery, which would be of little benefit to the foetus [23].

In our cohort, we could not find an increased rate of obstetric complications such as APH, PPH and uterine rupture; however, we found an increased rate of repeat caesarean section; around two thirds of the indication for caesarean sections in our cohort was a previous caesarean section.

The risk of repeated caesarean sections should be addressed in women with a previous uterine scar especially in anencephalic pregnancies as the risk of having a post-term pregnancy (prolonged gestation) is increased due to the hypothalamic-pituitary axis dysfunction [24]. In addition, induction of labour might not be a reliably safe option in women with a previous caesarean section due to an increased risk of uterine rupture [25–27].

Parents who plan for larger families should be counselled that the more caesarean sections a woman has the more risk would be anticipated for future pregnancies particularly massive obstetric haemorrhage and hysterectomy due to the increased risk of abnormal placentation [28–30].

Prenatal counselling should be therefore individualised based on the patient's obstetric history and parents' wishes for the current and future pregnancies.

Our study is the largest series to specifically address the maternal outcome when anencephalic fetuses are delivered after the age of viability. We would recommend performing an early fetal anatomy ultrasound, and thereby ensure that parents are adequately and timely counselled about their available options including the option of termination of pregnancy.

We would also recommend further studying the mode of delivery and the rate of caesarean section in similar cases as well as studying the future pregnancy outcomes of women with previous anencephalic pregnancies.

Author's statement

Conflict of interest: Authors state no conflict of interest.

Material and methods: Informed consent: Informed consent has been obtained from all individuals included in this study waiver of informed consent granted from the institutional research board.

Ethical approval: The research related to human subject use has complied with all the relevant national regulations, and institutional policies, and is in accordance with the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

Declaration: This article represents original work and has not been submitted for publication elsewhere.

References

[1] Lemire RJ, Beckwith JB, Warkuny J. Anencephaly. New York: Raven Press; 1978.
[2] Stone DH. The declining prevalence of anencephalus and spina bifida: its nature, causes and implications. Dev Med Child Neurol. 1987;29:541–6.
[3] Jones KL. Smith's recognizable patterns of human malformation. 6th ed. Philadelphia: WB Saunders; 2006. p. 704–5.
[4] Obeidi N, Russell N, Higgins JR, O'Donoghue K. The natural history of anencephaly. Prenat Diagn. 2010;30:357–60.
[5] Peabody JL, Emery JR, Ashwal S. Experience with anencephalic infants as prospective organ donors. N Engl J Med. 1989;321:344–50.
[6] McAbee G, Sherman J, Canas JA, Boxer H. Prolonged survival of two anencephalic infants. Am J Perinatol. 1993;10:175–7.
[7] Medical Task Force on A. The infant with anencephaly. N Engl J Med. 1990;322:669–74.
[8] Mathews TJ, Honein MA, Erickson JD. Spina bifida and anencephaly prevalence – United States, 1991–2001. MMWR Recomm Rep. 2002;51:9–11.
[9] Cook RJ, Erdman JN, Hevia M, Dickens BM. Prenatal management of anencephaly. Int J Gynaecol Obstet. 2008;102:304–8.

[10] Johnson SP, Sebire NJ, Snijders RJ, Tunkel S, Nicolaides KH. Ultrasound screening for anencephaly at 10–14 weeks of gestation. Ultrasound Obstet Gynecol. 1997;9:14–6.
[11] Cameron M, Moran P. Prenatal screening and diagnosis of neural tube defects. Prenat Diagn. 2009;29:402–11.
[12] Salomon LJ, Alfirevic Z, Berghella V, Bilardo C, Hernandez-Andrade E, Johnsen SL, et al. Practice guidelines for performance of the routine mid-trimester fetal ultrasound scan. Ultrasound Obstet Gynecol. 2011;37:116–26.
[13] Blaas HG. Detection of structural abnormalities in the first trimester using ultrasound. Best Pract Res Clin Obstet Gynaecol. 2014;28:341–53.
[14] Rossi AC, Prefumo F. Accuracy of ultrasonography at 11–14 weeks of gestation for detection of fetal structural anomalies: a systematic review. Obstet Gynecol. 2013;122:1160–7.
[15] Salomon LJ, Alfirevic Z, Bilardo CM, Chalouhi GE, Ghi T, Kagan KO, et al. ISUOG practice guidelines: performance of first-trimester fetal ultrasound scan. Ultrasound Obstet Gynecol. 2013;41:102–13.
[16] Souka AP, Pilalis A, Kavalakis Y, Kosmas Y, Antsaklis P, Antsaklis A. Assessment of fetal anatomy at the 11–14-week ultrasound examination. Ultrasound Obstet Gynecol. 2004;24:730–4.
[17] Van Mieghem T, Hindryckx A, Van Calsteren K. Early fetal anatomy screening: who, what, when and why? Curr Opin Obstet Gynecol. 2015;27:143–50.
[18] Chasen ST, Kalish RB. Can early ultrasound reduce the gestational age at abortion for fetal anomalies? Contraception. 2013;87:63–6.
[19] Levels M, Sluiter R, Need A. A review of abortion laws in Western-European countries. A cross-national comparison of legal developments between 1960 and 2010. Health Policy. 2014;118:95–104.
[20] Lawson HW, Frye A, Atrash HK, Smith JC, Shulman HB, Ramick M. Abortion mortality, United States, 1972 through 1987. Am J Obstet Gynecol. 1994;171:1365–72.
[21] Chakravarti S, Pai M, Kushtagi P. Contracted pelvis and cephalopelvic disproportion. Manual of obstetrics e-book. 4th ed. India: Elsevier Health Sciences; 2015. p. 335.
[22] Cammu H, Martens G, Ruyssinck G, Amy JJ. Outcome after elective labor induction in nulliparous women: a matched cohort study. Am J Obstet Gynecol. 2002;186:240–4.
[23] Chervenak FA, Farley MA, Walters L, Hobbins JC, Mahoney MJ. When is termination of pregnancy during the third trimester morally justifiable? N Engl J Med. 1984;310:501–4.
[24] Novy MJ, Walsh SW, Kittinger GW. Experimental fetal anencephaly in the rhesus monkey: effect on gestational length and fetal and maternal plasma steroids. J Clin Endocrinol Metab. 1977;45:1031–8.
[25] McDonagh MS, Osterweil P, Guise JM. The benefits and risks of inducing labour in patients with prior caesarean delivery: a systematic review. Br J Obstet Gynaecol. 2005;112:1007–15.
[26] Palatnik A, Grobman WA. Induction of labor versus expectant management for women with a prior cesarean delivery. Am J Obstet Gynecol. 2015;212:358.e1–6.
[27] Landon MB, Hauth JC, Leveno KJ, Spong CY, Leindecker S, Varner MW, et al. Maternal and perinatal outcomes associated with a trial of labor after prior cesarean delivery. N Engl J Med. 2004;351:2581–9.
[28] Miller DA, Chollet JA, Goodwin TM. Clinical risk factors for placenta previa-placenta accreta. Am J Obstet Gynecol. 1997;177:210–4.
[29] Clark SL, Koonings PP, Phelan JP. Placenta previa/accreta and prior cesarean section. Obstet Gynecol. 1985;66:89–92.
[30] Silver RM, Landon MB, Rouse DJ, Leveno KJ, Spong CY, Thom EA, et al. Maternal morbidity associated with multiple repeat cesarean deliveries. Obstet Gynecol. 2006;107:1226–32.

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