Anencephaly: knowledge and opinion of gynecologists, obstetricians and pediatricians in Goiânia

Marcus Vinícius Martins de Castro Santana [1], *Fernanda Margonari Cabral Canêdo* [2], *Ana Paula Vecchi* [3]

Abstract

In order to analyze the knowledge and opinion of physicians about anencephaly, a cross-sectional study was performed, including 70 obstetrician-gynecologists and pediatricians of two hospitals in Goiania, Brazil. The interviewees answered a survey of 20 true or false closed questions. Their opinions were evaluated through a 5-subject questionnaire, with 31 affirmations with a Likert-type response scale. Most of the interviewees (70%) affirmed to have attended classes on anencephaly. The average of correct answers was 13.17. The assertive questions with the highest percentage of correct answers were on anencephalic pregnancy (80%) and the distinction between anencephaly and brain death (72%). The questions with the lowest number of correct answers were about the donation of anencephalic born alive babies' organs (35%) and about the legislation that permits pregnancy termination when anencephaly had been unequivocally diagnosed (47.1%). Among those heard, 30.41% agreed on the fact that anencephalic babies have life. In conclusion, anencephaly is still a controversial topic and physicians need to acquire more knowledge on the subject.

Keywords: Abortion. Anencephaly. Brain death. Life. Congenital abnormalities. Embryonic development.

Resumo

Anencefalia: conhecimento e opinião dos médicos ginecologistas-obstetras e pediatras de Goiânia

Objetivando analisar o grau de conhecimento e opinião dos médicos sobre anencefalia, foi realizado estudo transversal com 70 ginecologistas-obstetras e pediatras de dois hospitais de Goiânia. Os entrevistados responderam a questionário com 20 perguntas fechadas, com opções "verdadeiro" ou "falso", abrangendo cinco temas distribuídos em 31 afirmações com respostas em escala tipo Likert. A maioria dos entrevistados afirmou ter assistido a aula sobre anencefalia (70%), e a média de acertos foi de 13,17 questões. As afirmações com mais acertos versavam sobre a gestação do anencéfalo (80%) e que anencefalia não significa morte encefálica (72%). As questões com menor número de acertos abordavam doação de órgãos de anencéfalo nascido vivo (35%) e a legislação que permite a interrupção da gestação perante diagnóstico inequívoco de anencefalia (47,1%). Dos profissionais ouvidos, 30,41% concordaram que o anencéfalo tem vida. Conclui-se que a anencefalia ainda é tema polêmico e necessita ser mais conhecida entre médicos.

Palavras-chave: Aborto. Anencefalia. Morte encefálica. Vida. Anormalidades congênitas. Desenvolvimento embrionário.

Resumen

Anencefalia: conocimiento y opinión de los ginecólogos, obstetras y pediatras en Goiânia

Con el objetivo de analizar el grado de conocimiento y opinión de los médicos sobre la anencefalia, fué hecho estudio transversal con 70 médicos ginecólogos/obstetras y pediatras de los hospitales de Goiânia, Brasil. Los entrevistados respondieron un cuestionario de 20 preguntas de verdadero o falso, y se evaluaron sus opiniones a través de un cuestionario con cinco preguntas que contenían 31 opciones de respuestas en escala tipo *Likert.* La mayoría (70%) participó en clases sobre anencefalia. El promedio de respuestas correctas fue del 13. 17 preguntas. Las afirmaciones con mayor número de respuestas correctas se registraron en relación a la gestación del anencéfalo (80%) y sobre que no existe en este caso muerte encefálica (72%). Las preguntas con menores números de respuestas correctas se relacionaron a la donación de órganos del anencéfalo nacido vivo (35%) y sobre la legislación que permite la interrupción de la gestación frente al diagnóstico inequívoco de anencefalia (47,1%). El 30,41% estuvo de acuerdo que el anencéfalo tiene vida. A la conclusión se verfiicó que la anencefalia es aún un tema polémico y necesita un mayor conocimiento entre los médicos.

Palabras clave: Aborto. Anencefalia. Muerte encefálica. Vida. Anomalías congénitas. Desarrollo embrionario.

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1. Graduando kaisso@hotmail.com **2. Graduanda** femargonari@hotmail.com **3. Doutora** anapaulavecchi@gmail.com – Pontifícia Universidade Católica de Goiás (PUC), Goiânia/GO, Brasil.

Correspondência
Ana Paula Vecchi – Rua Madri, 22, quadra 17, lote 20, Jardins Madri CEP 74369-100. Goiânia/GO, Brasil.

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Research articles

Congenital malformations lead to morbidity and mortality, and the neural tube defects (NTD) are their second most common cause, affecting 0.5 to 2 pregnancies In every 1,000 [1]. According to the Federação Brasileira das Associações de Ginecologia e Obstetrícia - Febrasgo (Brazilian Federation of Gynecology and Obstetric Associations), there are no reliable data in Brazil about the actual incidence of spina bifida, myelomeningocele and other NTD (anencephaly and encephalocele), which vary in the different regions of the country. In the Northeast region there reports of specific area series in a tertiary center in which the prevalence was of five cases in every thousand births. In the South, a 14 year population analysis found a prevalence of about six cases per 10 thousand births and, in the Southeast, the prevalence was of 1.13 per thousand born alive [2].

In humans, the most common NTD are anencephaly, consisting in the failure in the closing of the anterior neural tube, and myelomeningocele, also characterized by imperfection in the closing of the neural tube, but in the caudal end, culminating in spine defects [3]. Anencephaly may be divided in meroanencephaly, when cranial bones and the anterior brain are affected and some vegetative functions are preserved [1,4], and holonencephaly with failure in closing of the posterior part of the brain and cranial bones. It is evident that anencephaly presents varieties with severity depending on the degree of involvement [1].

Anencephaly would be better defined as the failure to close the neural tube between the third and fourth weeks of pregnancy (between the 23rd and the 26th day of the embryo), resulting in total or partial absence of the cranial vault (skull and scalp) and brain [3,5]. In fact, although the term "anencephaly" (from the Greek *an*, "without", and *enkephalos*, "encephalon") is commonly used, it is a misleading term, since the encephalon – which by anatomic definition is the part of the central nervous system located in the cranium, made up of the brain (telencephalon and diencephalon), cerebellum and brainstem (midbrain, pons and medulla oblongata) [6] – is not completely absent [7], as the brainstem, cerebellum, diencephalon and parts of the telencephalon are usually present.

The exact etiology of anencephaly and other NTD, like myelomeningocele or spina bifida, is still unknown. However, environmental conditions such as socioeconomic status and genetic influences (mutation of methylenetetrahydrofolate reductase; changes in microRNA regulation, via mitogen-activated protein kinase - MAK) are known as some of the factors that can lead to these conditions [5,8-12].

Among the environmental factors, the deficiency in folic acid during pregnancy stands out. Randomized clinical trials and several observational studies showed that 50% to 85% of the cases of NTD can be prevented if women receive folic acid supplementation at a daily dose of 0.4 mg daily in the periconceptional period [2,13]. Considering the low adherence to medicine supplementation by the population, and in order to reduce the prevalence of maternal anemia and neural tube defects, the Brazilian government established the mandatory fortification of wheat and maize flour with iron and folic acid, which was effectively implemented in the country from June 2004 [14].

Most anencephalic newborns die within days or weeks after birth [15]. In some rare cases, they survive longer, as was the case, at the Hospital Materno Infantil de Goiânia (Mother and Child Hospital of Goiania) , of Samuel, who lived six months; or Vitória de Cristo, a nationally known episode, who survived two years and six months; or, still, in Ribeirão Preto, of a three years and seven moths old girl, published in 2011 [4].

Machado et al. [8] performed a retrospective assessment of 108 pregnancies of anencephalic fetuses in a ten-year period. In the group in which the parents decided to keep the pregnancy until the end (n = 53), 62% of the fetuses were born alive and 38% died in the uterus. Of the 31 born alive, 94% died in the first 24 hours, 67% of which passed away in the first hour. The authors did not inform if there was advanced support to these newborns.

The reaction of couples facing the prenatal diagnosis of a fetal abnormality may involve feelings of anger, hopelessness, inadequacy and sleep and eating disorders, negative feelings that may be directed toward the health team, demonstrating that knowledge and understanding of professionals are essential to help the couple to adapt to the new reality [16,17]. The diagnosis of fetal malformation can have great impact on the self-esteem of parents, who may feel "defective" or incapable of generating a healthy child, and the feeling of guilt and responsibility for the malformation is very common [18].

Therefore, physicians responsible for the diagnosis of fetal malformation are instruments of aid and support to the families of malformed fetuses and, being charged with the *therapeutic anticipation of delivery* when there is mutual consent, are capable of influencing the decision of the parents with their knowledge and personal conceptions. Thus, the

investigation of the degree of knowledge and the opinion of physicians about anencephaly is fundamental for the analysis of the methods used and the guidance provided to women pregnant of anencephalic fetuses.

Methods

This crosscut study was carried out at the Hospital Materno Infantil (Mother and Child Hospital) and at the Santa Casa de Misericórdia de Goiânia (Saint Goiania Mercy House) (SCMG), in the period form January to May, 2015, with the use of a self-applied questionnaire with 36 questions divided in three sections: 1) personal data, such as age, sex, ethnic group, income, profession, religion and religiosity; 2) general knowledge about anencephaly; and 3) opinions about anencephaly.

The first part consisted in identification data, Including the DUREL scale (Duke University Religion Index, Brazilian version), which checks for three of the main aspects of the relationship between religiosity and health outcomes, through five items: the first two elements address aspects of organizational religious activity (ORA), related to the degree of attendance to religious meetings (going to churches, temples or ceremonies for example), and non-organizational religious activity (NORA), which measures the frequency with which the interviewee performs individual religious activities (prayer, Bible study, devotional activities etc.). Both indicators were reproduced from epidemiological studies from the United States [19] and associated to indices of physical and mental health, and social support. The three last items comprehend the dimension of intrinsic religiosity (IR), which refers to the attempt to experience religiosity in all dimensions of life and were extracted from Hoge's IR scale, according to the their adequacy to the total score of this scale and including outcomes in health and social support. Moreira et al. recommend that, in the analysis of the DUREL results, the scores of the three dimensions (ORA, NORA and IR) must not be added but, instead, that they be analyzed separately [20,21].

For the present study, we used the definitions of religion, religiosity and spirituality by Koenig et al. [19] who state : 1) *religion is an organized system of beliefs, practices, rituals and symbols designed to facilitate the access to the sacred (God, major power, supreme truth)*; 2) *religiosity refers to how much and individual believes in, follows and practices a certain religion* and; 3) *spirituality refers to a personal search for understanding related to larger existential questions (for example, the end and the meaning of life) and its relationships with the sacred and/or transcendent*. This process does no necessarily result in the development of practices or in the creation of religious communities [19-21].

The second part of the questionnaire consisted in 20 questions of the true or false type, the value 1 being attributed to correct answers and 0 being attributed to incorrect answers, in such a way that the score ranged from 0 to 20. The third part of the questionnaire identified the opinion of the interviewees through 31 statements in a Likert-type scale, in which they marked the options: "agree very much", "partially agree", "do not agree or disagree", "partially disagree", or "disagree very much".

Questionnaires were previously applied to a group of ten students in the the sixth year of medical school, whose role was to criticize the formulation of questions, in order to test the clarity of the language and the objectivity of the instrument.

Inclusion criteria for the study were: to be a gynecologist-obstetrician and pediatrician at the HMI or the SCMG; to be a resident physician in pediatrics or gynecology at the HMI or the SCMG who accepted to participate in the study and sign the free and informed consent form. Questionnaires with incomplete or inadequate answers were not used in the analysis. All participants signed the free informed consent form.

The sample of 155 physicians, including residents, was calculated through the Epinfo 7 software, adopting a 5% sample error, expected frequency of 50% and 95% confidence interval.

Results

In total, 155 questionnaires were distributed but only 70 physicians filled them correctly and were included in the study. Of the remaining, 35 did not want to answer, 28 did not return the questionnaire and 22 answered in incomplete or inadequate ways, marking more than one alternative.

Questionnaires were answered in about ten minutes in the presence of the researcher, at the workplace of the physicians, after information on the study and signature of the consent form for participation.

Most physicians were female (76%), white (70%), catholic (53%) and attended class on anencephaly (70%). The average ORA score in the DUREL scale was 3.21 (1-6); for NORA it was 2.81 (1-6); and for IR, 4.9 (1-15). The average number of correct answers was 13.17 questions, of the total 20 (Table 1).

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Table 1. General characteristics of physicians

	n	% (n = 70)
Sex		
male	17	24.3
female	53	75.7
Color/ethnic group		
white	49	70
black	1	1.4
brown	17	24.3
yellow	3	4.3
Indian	–	0
Religion		
catholic	37	52.9
protestant/evangelical	11	15.7
spiritist	16	22.9
other	2	2.8
none	4	5.7
Specialty		
gynecologist-obstetrician (GO)	6	8.6
pediatrician	28	40
resident in pediatrics	14	20
resident in GO	22	31.4
Had classes on anencephaly	49	70
Age (average)	34	
Total of correct answers (average)	13.17	

The most significant statements to measure the knowledge on the matter referred to survival rates (77.1%) and gestation of the anencephalic fetus (80%), brain death criteria (87.1%) and that anencephaly does not mean brain death (72.8%). Questions with the lowest number of correct answers addressed the donation of organs of the anencephalous born alive (35.7%) and the law allowing the interruption of pregnancy after the unequivocal diagnosis of anencephaly (44.3%). These data can be seen in Table 2.

Concerning the opinion of physicians, 47.14% agreed very much that the anencephalous has life, but 41.71% disagree very much that the anticipation of delivery is an abortion. Of the professionals interviewed, 45.71% (sum of the values of the ones who agree partially and the ones who agree very much) suggest therapeutic anticipation to avoid parental suffering and 88.57% (sum of the values of the ones who agree partially and the ones who agree very much) refer these parents to psychological help. When the parents opt for the therapeutic interruption, 15.71% of the physicians agree very much to interrupt the pregnancy without a problem; 15.71% of them would interrupt but would feel uncomfortable; 27.14% would only proceed under a court order and 32.86% would refer the case to another physician. In the care for the anencephalic newborn, 48.57% agree very much that reanimation must be complete and 7.14% consider that nothing should be done. 65.71% disagree very much or partially with the abortion before 12 weeks, but 50% agree very much or partially with the abortion of anencephalous, as shown in Table 3.

Discussion

Anencephaly is one of the most common malformations of the neural tube and results in the total or partial absence of the cranial vault (skull ans scalp) and the brain [3,5], but not the whole encephalon, as frequently divulged outside the academic environment. The score above 60% obtained by physicians as to its definition reflects their knowledge of encephalon anatomy and function, certainly acquired during their academic training.

As to the survival rates, their score was 77%, showing that most had previous access to the matter – in fact, 70% pf the physicians declared to have had class on anencephaly. However, less than half of them demonstrated knowledge of the norms of the Conselho Federal de Medicina (Brazilian Federal Medicine Council - CFM) and the Supremo Tribunal Federal - STF (Brazilian Supreme Court) relating to anencephalic pregnancy.

About this subject, on April 12th 2012, the STF judged upheld the Arguição de Descumprimento do Preceito Federal-54" ADPF-54 [22] (Claim of Breach of Federal Precept-54) and decided that the therapeutic anticipation of delivery of anencephalic fetuses is not a crime of abortion predicted in the Brazilian Penal Code, thus, not requiring previous authorization. On May 10th 2012, the CFM issued Resolution 1,989/2012 [23], which, in article 1, defines that *in the occurrence of unequivocal diagnosis of anencephaly the physician may, at the request of the pregnant woman, independent of State authorization, interrupt the pregnancy*. Article 2 defines that:

the diagnosis of anencephaly is done by ultrasonography examination performed from the 12th (twelfth) week of gestation and must contain:

I – two photos, identified and dated: one with the face of the fetus in sagittal position; the other, with the view of the fetal head in cross-section, showing the absence of the skull and identifiable brain parenchyma;

II – report signed by two physicians trained for such diagnosis [23].

Table 2. Knowledge of the physicians

	Answer	Correct	
		n	% n = 70
Knowledge of physicians about the definition and survival of anencephalic fetuses			
1. Anencephaly is defined as the total absence of the encephalon	F	45	64.3
2. Anencephaly is defined as the total absence of the brain and midbrain	F	46	65.7
3. Anencephaly is defined as a failure in closing the neural tube between the third and fourth week of gestation (23rd and 26th day of the embryo), resulting in the total or partial absence of the cranial vault (skull and scalp) and the brain.	T	41	58.5
4. Considering the anatomic definition, the term "anencephaly" is wrong.	T	59	84.3
5. The structures of the central nervous system that are generally present in the anencephalic fetus are: diencephalon, midbrain, pons, medulla oblongata and cerebellum	T	37	52.8
6. Most anencephalic fetuses die in the uterus.	F	36	51.4
7. Most anencephalic fetuses die in the first day, and only 5% survive longer than six days.	T	54	77.1
Knowledge of physicians about brain death, anencephaly and donation of organs			
8. Anencephaly may be considered as brain death.	F	51	72.8
9. In Brazil, criteria for brain death follow CFM Resolution 1,480/1997, defined by the irreversible clinical state in which brain and brain stem functions are irreparably compromised.	T	61	87.1
10. We can asses the functioning of the brainstem through pupillary, corneal, vestibular-caloric, oculocephalic and cough reflexes, and apnea test.	T	63	90
11. In cephalic death, brainstem reflexes are absent.	T	55	78.6
12. In the anencephalous, brainstem reflexes are present.	T	49	70
13. The immaturity of the central nervous system of the child determines stricter criteria for determining brain death. The diagnosis is only possible after the seventh day of life.	T	28	40
14. According to the American Academy of Pediatrics, anencephalic children can be donors of vital organs.	F	25	35.7
Knowledge of physicians about the gestation of anencephalic fetuses and the legislation			
15. The gestation of the anencephalic fetus requires special monitoring, and the most frequently reported complications are polyhydramnios and prematurity	T	56	80
16. Pregnancy is complicated by hyperemesis in most cases.	F	39	55.7
17. The gestation of the anencephalic fetus endangers the woman's life	F	53	75.7
18. In the event the unequivocal diagnosis of anencephaly, the physician may terminate the pregnancy at the request of the pregnant woman, regardless of State authorization.	T	31	44.3
19. In the event the unequivocal diagnosis of anencephaly, the physician may terminate the pregnancy at the request of the pregnant woman provided there is authorization by the State.	F	33	47.1
20. In the event the unequivocal diagnosis of anencephaly, the physician may not terminate the pregnancy.	F	54	77.1

Ultrasonography is an effective modality for the prenatal diagnosis of the NTD [24]. The total understanding of the normal ultrasonographic appearance of the central nervous system at different gestational ages is crucial for the precise diagnosis because the absence of a structure must be considered as normal or abnormal, depending on the gestational age [25]. Camano et al. [26] studied 5,351 ultrasonographies with measurement of nuchal translucency, calculated the risk of chromosomal anomalies and monitored these patients. Of these, 193 (3.6%) of the exams showed alterations, with the risk of anomalies above 1:300. of the total newborns, 84.8% did not present chromosomal anomalies or malformations diagnosed during their stay in the nursery.

In a populational based study of 55,226 pregnancies [25], of 143 newborns with defects in the central nervous system (CNS), 85 had prenatal ultrasound performed between the 16th and 20th weeks of gestation. The diagnosis of CNS alterations during the prenatal was done in 64 fetuses (75%), was not done in 17 (20%) and was questionable in 4 (5%). Short exam time, instead of low sensitivity, was the most important factor for the detection of NTD, since it is known that exam sensitivity may reach 100% when performed by trained professionals.

Table 3. Opinions of physicians about anencephaly (Likert scale)

	Disagree very much		Partially disagree		Do not agree or disagree		Partially agree		Agree very much			TOTAL %
	n	%	n	%	n	%	n	%	n	%		
1. General statements on anencephaly												
a) The anencephalic fetus has life	8	11.43%	5	7.14%	9	12.86%	15	21.43%	33	47.14%	70	100.00%
b) Anencephaly is an anomaly that is incompatible with life	6	8.57%	7	10.00%	1	1.43%	11	15.71%	45	64.29%	70	100.00%
c) There may be consciousness in the anencephalic fetus	32	45.71%	8	11.43%	10	14.29%	18	25.71%	2	2.86%	70	100.00%
d) I consider the anencephalic fetus as in brain death	12	17.14%	17	24.29%	8	11.43%	16	22.86%	17	24.29%	70	100.00%
e) I am in favor of the donation of the organs of the anencephalic fetus	8	11.43%	4	5.71%	3	4.29%	11	15.71%	44	62.86%	70	100.00%
f) I consider the therapeutic anticipation of delivery of the anencephalic fetus as an abortion	29	41.43%	3	4.29%	9	12.86%	14	20.00%	15	21.43%	70	100.00%
2. In finding the diagnosis of anencephaly, how do you advise the mother?												
a) I suggest the therapeutic anticipation of delivery, since the anomaly is incompatible with life	27	38.57%	5	7.14%	10	14.29%	8	11.43%	20	28.57%	70	100.00%
b) I suggest the therapeutic anticipation of delivery to avoid parental suffering	20	28.57%	8	11.43%	10	14.29%	11	15.71%	21	30.00%	70	100.00%
c) I talk to the parents and let them decide	4	5.71%	3	4.29%	9	12.86%	11	15.71%	43	61.43%	70	100.00%
d) I refer them to psychological support only if they do not accept the anticipation of delivery	42	60.00%	8	11.43%	5	7.14%	6	8.57%	9	12.86%	70	100.00%
e) I always refer them to psychological support	3	4.29%	2	2.86%	3	4.29%	4	5.71%	58	82.86%	70	100.00%
f) I ask the religion of the parents and try to direct the decision according to their conception	16	22.86%	9	12.86%	16	22.86%	14	20.00%	15	21.43%	70	100.00%
g) I have difficulties talking about the diagnosis	16	22.86%	9	12.86%	10	14.29%	19	27.14%	16	22.86%	70	100.00%
3. When the parents decide for the abortion of the anencephalic fetus, you:												
a) Proceed without any problem	33	47.14%	5	7.14%	9	12.86%	12	17.14%	11	15.71%	70	100.00%
b) Proceed, but feel bad about it	27	38.57%	7	10.00%	9	12.86%	16	22.86%	11	15.71%	70	100.00%
c) Proceed, but only under a court order	30	42.86%	5	7.14%	8	11.43%	8	11.43%	19	27.14%	70	100.00%
d) Refer the case to another physician	20	28.57%	6	8.57%	16	22.86%	5	7.14%	23	32.86%	70	100.00%
4. When faced with a live newborn in the delivery room, you:												
a) Proceed with the standard care norms for newborns, including reanimation and advanced support (mechanical ventilation and vasoactive drugs)	20	28.57%	5	7.14%	7	10.00%	4	5.71%	34	48.57%	70	100.00%
b) Proceed with the standard care norms and, if necessary, use nasal oxygen	13	18.57%	4	5.71%	1	1.43%	10	14.29%	42	60.00%	70	100.00%
c) Proceed with the standard care norms: cry the newborn and place him/her in a warm crib. Do not use oxygen,	31	44.29%	6	8.57%	5	7.14%	10	14.29%	18	25.71%	70	100.00%
d) Do nothing	60	85.71%	1	1.43%	3	4.29%	1	1.43%	5	7.14%	70	100.00%
5. Are you in favor of abortion?												
a) In fetuses with less than 12 weeks	39	55.71%	7	10.00%	6	8.57%	8	11.43%	10	14.29%	70	100.00%
b) In case of rape	23	32.86%	7	10.00%	5	7.14%	6	8.57%	29	41.43%	70	100.00%
c) In case of anencephaly	24	34.29%	6	8.57%	5	7.14%	10	14.29%	25	35.71%	70	100.00%
d) In case of severe malformations with little chances of living	28	40.00%	9	12.86%	5	7.14%	8	11.43%	20	28.57%	70	100.00%
e) In any malformation	47	67.14%	10	14.29%	9	12.86%	3	4.29%	1	1.43%	70	100.00%
f) In adolescent mothers	64	91.43%	5	7.14%	0	0.00%	1	1.43%	0	0.00%	70	100.00%
g) In low-income mothers	66	94.29%	3	4.29%	1	1.43%	0	0.00%	0	0.00%	70	100.00%
h) When the mother does not want the child	59	84.29%	4	5.71%	1	1.43%	2	2.86%	4	5.71%	70	100.00%
i) When the mother's life is at risk	23	32.86%	4	5.71%	10	14.29%	13	18.57%	20	28.57%	70	100.00%
j) Never	31	44.29%	7	10.00%	12	17.14%	4	5.71%	16	22.86%	70	100.00%

Although fast magnetic resonance imaging (fast MRI) can also be used to reproduce the image of the fetus, ultrasonography is still the mode of election in the evaluation of fetuses in risk of NTD, given its high rate of detection [25]. The dose alpha-fetoprotein (AFP) may also be useful in the diagnostic suspicion of meroanencephaly, since a high AFP level in the amniotic fluid may suggest NTD [27].

The great majority of interviewees (80%) were correct in that the most common complications of the gestation are polyhydramnios and prematurity [8,28,29]. Machado et al. [8] found prematurity as the most common complication in 83% of the cases analyzed, and polyhydramnios in 15%, against 39% reported in other studies. Obeidi et al. [29] studied 26 uninterrupted pregnancies of anencephalous, of which 7 (27%) were complicated by polyhydramnios and 4 deliveries (15%) were complicated by shoulder dystocia. Average duration of gestation to delivery was 35 weeks (range: 22-42); 69% of labors were induced to an average gestation of 34 weeks. Polyhydramnios may lead to respiratory discomfort in the woman, but does not bring risk to life.

Study involving 456 anencephalous found 12.7% of associated malformations, markedly cleft lip and / or clef palate and omphalocele. Hypoplasia of the adrenal glands and intestinal aganglionosis were frequent [24]. Machado et al. [8] demonstrated association of at least one other malformation in 39% of anencephalic newborns and fetuses, the most frequent being facial and renal defects. Association to other cardiac, pulmonary and skeletal malformations are less common [30-32].

Differences between brain death and anencephaly

In Brazil, the criteria for brain death, equivalent to clinical death and represents the irreversible state in which brain functions (telencephalon and diencephalon) and brain stem functions are irreparably compromised [33], follow CFM resolution 1,480/1997 [34], as informed by Ribas. The diagnosis of death brain must be established after two clinical examinations performed by different professionals not linked to the transplant team, with minimal interval of six hours, and the performance of complementary exam showing the absence of electrical activity in the brain or the absence of brain perfusion or metabolism is also mandatory [33-35].

The immaturity of the infant's central nervous system determines stricter criteria for determining brain death, for which the diagnosis is only possible after the seventh day of life. Newborns included in the age group ranging from seven days to two months of age need an interval of at least 24 hours between clinical exams besides in addition to the completion of two electroencephalograms (EEG) [33].

The term "brain death" is inadequate, for the brain comprehends the telencephalon and the diencephalon, not including the brainstem, and the complete dysfunction of the brainstem is *sine qua non* condition for the diagnosis of brain death. The presence of brainstem functions can be assessed through pupillary, corneal, vestibular-caloric, oculocephalic and cough reflexes, and the apnea test. All these reflexes will be absent in brain death [33].

The anencephalous, by definition, shows the absence of the brain and vault, but the cerebellum and brainstem are present, as well as their reflexes [15]. Live-born anencephalous babies show functions of the brainstem, with spontaneous breathing and, frequently, some reflex responses, such as suction [24,36]. Therefore, there is an evident difference between the anencephalous and the patient in brain death: the functioning of the brainstem. Therefore, the anencephalous cannot be considered dead [37], except when there is cardiac arrest. The physicians interviewed show high rate of correct answers (above 72.8%) about the definitions of brain death and its differences is relation to the anencephalous.

Despite the fact that most interviewees were pediatricians or residents in pediatrics, the question on the possibility of the donation of organs from anencephalic babies was the one with the lowest number of correct answers, only 35.7 of the physicians answered that anencephalic babies cannot be donors. This is the position, since 1992, of the American Academy of Pediatrics, since these newborns do not fulfill the criteria for brain death – the brainstem keeps working, showing spontaneous breathing and heartbeat and, when there is cardiac and respiratory arrest, the organs are already in the process of ischemia [15].

Opinions of physicians and psychological aspects

There was a tendency among physicians to consider the anencephalous a living being but incompatible with life, probably due to its short survival. This is reflected in the conduct adopted facing an anencephalous newborn, for the majority of the physicians agreed very much to proceed with the standard care norms to the newborn, including reanimation and advanced support.

Although those who survived labor show spontaneous breathing, most authors postulate the

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absence of consciousness as a result of the non-functioning of the brain. Funayama et al. [4] described the physical exam of a patient with merocrania, a type of anencephaly, who presented which had quadriplegia, global hypotonia with occasional body hypertonia in a decorticated posture, hyperreflexia, ankle clonus and plantar extensor response. The infant had complete absence of the cerebrum and rudimentary brain stem on the magnetic resonance imaging, but was able to suck, respond to pain with muscle contractions or weak cry, had complete closure of the eyelids, good apposition of the lips and did not present sialorrhoea.

The anencephalous child also moved her head and eyes towards sound and was able to smile to sound and tactile stimuli. Smiling has been recognized in anencephalic infants and was related to REM (Rapid Eye Movement) sleep, that is anatomically correlated to the pons [4]. The smile in moments of pleasure and the look towards stimuli do not seem to be reflexes and Funayama et al. raise the possibility of a consciousness, even in the absence of the cerebral cortex, perhaps via brainstem [4].

A significant number of physicians suggest the therapeutic anticipation of delivery to avoid parental suffering, but they consider the decision on pregnancy interruption to be up to the couple. The large majority always suggest psychological monitoring, but asking about the religion of the parents with its consequent impacts is not common. The interviewees tended to be against abortion, except in the case of risk to the mother's life, rape and severe malformations with few chances of survival. Most physicians were catholic, a religion that is not favorable to abortion nor to therapeutic anticipation of delivery in case of anencephaly. The average value of organizational religious activity (ORA) was 3.21 (range: 1-6); that of non-organizational religious activity (NORA), was 2.81 (range: 1-6); and that of intrinsic religiosity (IR) was 4.9 (range: 1-15). We used the version validated by Moreira et al. [20], in which lower indices point to higher religiosity. For example, when assessing ORA, if the person goes to the temple more than once a week, she receives a score of 1, whereas the one who answers "never" receives a score of 6. The same logical sequence is followed for the question that assesses non-organizational religious activity: *How often do you spend time in private religious activities, such as prayer, meditation or Bible study (or study of other religious texts)?* And, lastly, in the assessment of intrinsic religiosity, the interviewee answers three questions with five possible answers that can be added and for which the lowest result, likewise, represents higher religiosity.

Benute et al. [16] monitored 35 pregnant women with diagnosis of lethal fetal malformation and showed that 60% of them showed negative feelings (shock, anguish, sadness, resignation, plans destruction, outrage, fear, shame, worthlessness, crying, inability to be a mother, indignation and insecurity as a woman) during the period they had to decide for or against the interruption of pregnancy. After the interruption of pregnancy, 62.8% of the patients reported memories of the imaginary child or negative feelings, although 91% stated that they would repeat the decision in a similar situation and 60% would suggest abortion to another person in a similar situation [16].

It is extremely important to make sure the pregnant woman – or the couple – has adequately understood the diagnosis and the prognostics for the baby, survival time and possible risks of the pregnancy, and, whatever the decision of the pregnant woman, the physician must inform her of the consequences, including the risks resulting form each one. Both the woman who decides to keep the pregnancy and the one who chooses for its interruption must, if they wish so, receive assistance from the multi-professional team in places where this is available. Psychological monitoring is fundamental and helps the couple face reality and make the necessary possible decisions concerning, for example, issues like the interruption (or not) of the pregnancy, what will be done after the birth or passing away of the baby, if there will be some type of farewell ritual and how this will be. It is necessary, in many cases, to mediate the dialogue between the couple for these decisions to be made jointly, considering the beliefs, the feelings and the desires of both [17].

Cases in which the babies survived longer despite the severity of of the malformation and the initial prognosis are among the ones that cause the most controversy about legal interruption of pregnancy due to incompatible with life fetal malformations [18]. Some women pregnant of anencephalic fetuses decide to keep the pregnancy until the end even having the possibility of of having a legal interruption, and communicate affectionately with the baby during the gestational period. The conscience that they are capable of caring for their child the best possible way and of learning from this experience makes them feel gratified [16-18,33-35,37,38].

The way to deal with the situation is also related to family's religion and religiosity. According to Bortoletti et al. some evangelical pregnant

women *delegate all the process to the divine power and, despite submitting themselves to the medical procedures, are always 'in the wait' for God's 'miracle'* [39]. Catholic patients may also resort to similar measures, seeking support in prayer, spells or promises which may both revert the diagnosis and just comfort them and help them endure the suffering. Among spiritist pregnant women, the authors observed the predominance of the belief that the gestation has the function of allowing the baby *to be 'redeemed' of what she did in previous lives, putting the fetus in the position of 'debtor' and the mother in the place of one who has to go through this suffering to be redeemed form a possible 'karma'* [39], or even as an opportunity of growth and learning. If the religiosity of the parents interferes in facing the anencephalic condition of the child and in the decision about the interruption or not of the pregnancy, could the religiosity of the physician interfere in concepts and conducts facing a case of anencephaly?

When informing the diagnosis of fetal malformation, it is expected that the support to the patient be characterized by neutrality in the clarification on the diagnostic and therapeutic procedures. Thus, the impartiality of the professional is fundamental for the couple to turn to their own beliefs and convictions, unleashing a process of reflection that may help the eventual discussion on the interruption (or not) of the pregnancy. At any rate, these are issues that must be addressed during the appointments, evidently always respecting the desire of the couple [16,18].

This way, the knowledge and opinions of the physicians about anencephaly may help parents face this situation, as well as support them in any decision. Anencephaly is still a polemical issue and it more knowledge and discussion among physicians are necessary.

Final considerations

The diagnosis of anencephaly, a congenital malformation incompatible with life, accentuates the feelings of ambiguity of the healthy pregnancy, with predominance of negative thoughts of the couple, mainly the mother. It is fundamental that the medical team have knowledge about the prenatal diagnosis of anencephaly, of the legal provisions and, above all, the importance of the psychological monitoring of the couple, from the diagnosis until some months after the birth or interruption. The warm and neutral attitude of the physician, without presenting any opinion as absolute truth to the pregnant woman and the support of the entire multidisciplinary team allow the couple to make the decision according to their beliefs and to develop the arising mourning process. The choice is personal and must be respected by the professionals.

This study shows the importance of studying anencephaly and the consequent need for training of specialized teams – with physicians, psychologists and social assistants – to care for these pregnant women. The importance of psychological support to these professionals is also evident, since not all of them have the opportunity to reflect about their feelings and behaviors facing these cases, as well as on their own difficulties dealing with these issues. Thus, with the creation of specialized teams to deal with malformations, the medical team would become a great support for couples in facing and in overcoming this situation.

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Participation of the authors
The authors had equal participation in all phases of the study.

Annex

Frame 1. Duke University Religion Index (DUREL)

(1) How often do you attend church or other religious meetings?
1. More than once/week
2. Once a week
3. A few times a month
4. A few times a year
5. Once a year or less
6. Never

(2) How often do you spend time in private religious activities, such as prayer, meditation or Bible study (or study of other religious texts)?
1. More than once a day
2. Daily
3. Two or more times/week
4. Once a week
5. A few times a month
6. Rarely or never

The following section contains 3 statements about religious belief or experience. Please mark the extent to which each statement is true or not true for you.

(3) In my life, I experience the presence of the Divine (i.e., God or the Holy Spirit).
1. Definitely true of me
2. Tends to be true
3. Unsure
4. Tends not to be true;
5. Definitely not true;

(4) My religious beliefs are what really lie behind my whole approach to life.
1. Definitely true of me
2. Tends to be true
3. Unsure
4. Tends not to be true;
5. Definitely not true

(5) I try hard to carry my religion over into all other dealings in life.
1. Definitely true of me
2. Tends to be true
3. Unsure
4. Tends not to be true;
5. Definitely not true;

Source: Moreira-Almeida et al. Rev Psiquiatr. Clín. 2008;35(1);31-2. (Translator's note: English version based on Koenig and Büssing. Religions 2010, 1, 78-85.)