Is counselling for CCAM that difficult? Learning from parental experience

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Summary

Objective: **Cystic adenomatoid malformations (CCAM) are relatively rare developmental abnormalities of the lung. Despite outcome is usually benign, parents often exhibit high anxiety level. The purpose of the present study was to collect parents'subjective experience of communication of diagnosis when expecting a fetus with CCAM.**
Methods: **In the period ranging between 2004 to 2007 all couples referred to our hospital for prenatal suspicion of CCAM, were contacted to participate in the study by means of a questionnaire.**
Results: **Twenty-seven couples returned fully completed questionnaire and form the object of this study. 13/27 couples were recommended pregnancy termination. Major risks presented were: fetal distress (27/27), intrauterine death (16/27), death at birth (12/27). The most distressing moments were: communication of diagnosis and the time lag between communication of diagnosis and consultation with the surgeon.**
Conclusions: **Despite CCAM carries a favourable prognosis, couples often appear to receive incomplete information with risk of fetal distress and demise prevailing over survival rate and long term outcome. To provide the couple with the most accurate information concerning the anomaly and the associated prognosis to make informed decision a referral to a tertiary pediatric surgery unit should be made as soon as possible.**

Key words: antenatal counselling, prenatal diagnosis, CCAM, communication of diagnosis.

Introduction

Cystic adenomatoid malformations (CCAM) are relatively rare developmental abnormalities of the lung: they are mostly unilateral involving one lobe of the lung (1,2): outcome is usually benign. There is little research documenting what gynaecologists do when they discover this malformation and disclose the diagnosis to the couple, nor is there a standard method of practice in prenatal clinic setting. Little research has been done investigating parents's subjective experience when a CCAM is diagnosed in their fetus (3-5).
This study was designed to investigate parents' subjective experience of communication of diagnosis when expecting a fetus with CCAM and assess parental feelings soon after communication of diagnosis.

Material and Methods

In the period ranging from 2004 to 2007, 35 newborn infants were surgically treated at our Institution after prenatally detected CCAM. Couples were referred to our service for counselling after diagnosis.
After surgical correction of CCAM all couples were contacted to offer them possibility of participating in the present study. Couples who agreed to participate were sent a questionnaire made up of 11 items regarding the following aspects: main information perceived by parents at communication of diagnosis and parents'concerns. Questions were developed according to steps of the adaptation process (6, 7) as well as to interviews realized in the last 10 years with couples receiving antenatal diagnosis of CCAM. **The study was IRB approved.**

Results

A total of 35 couples with prenatal diagnosis of CCAM confirmed at birth were seen over the considered period and received the questionnaire. Twenty-seven (77%) couples returned it fully completed and form the object of this study. No cases of intrauterine death were observed neither were there couples opting for termination of pregnancy. In 9 cases cystis increased in size up to 28 weeks without causing any hydrops. In 5 cases cysts apparently disappeared

before birth but their persistence was confirmed by CT scan performed postnatally according to our surveillance policy. 2/27 children were operated on at birth because of symptoms. Time span between diagnosis and request to fill in questionnaire ranged from 4 to 8 months.
Educational level ranged from high school to college. Marital status was the following: married 19/27, cohabitant 8/27. Parity: 21 nullipara, 6 multipara. Maternal age ranged from 22 to 38 years, paternal age from 25 to 44 years. Parents were all Caucasian. Median gestational age at diagnosis was 21 week (range 16 to 32 weeks). In 27/27 couples communication of diagnosis was made by the sonologist. 8/27 couples reported having received information about quality of life after surgery, 10/27 couples regarding survival rate in CCAM patients. Parents reported they were presented the following risks: fetal distress 27/27, intrauterine death 16/27, stillbirth 12/27. At first announcement of the anomaly pregnancy termination was recommended by the gynaecologist performing ultrasound to 14 couples. 19/27 couples were referred to pediatric surgeon after diagnosis by the obstetrician. In 15 (55%) mothers and 16 fathers (59%) major concern after communication of diagnosis was of losing their baby during pregnancy. Main emotions since pregnancy were disorientation which occurred in 18 mothers (66%) and 20 fathers (74%) and fear in 8 (30%)mothers and 3 fathers (11%). The most distressing moments experienced by parents were: communication of diagnosis and time from communication of diagnosis to consultation with surgeon. Data obtained from questionnaire are reported in Table I.

Table 1 - Questionnaire administered to the couples.

Question Items Answer	Mother	Father
1) When was diagnosis first established?		
16-21wks	11	11
22-27wks	16	16
28-32wks	0	0
>33wks	0	0
2) Diagnosis was communicated by?		
Obstetrician	27	27
Other	0	0
3) Did the obstetrician convey information about quality of life?		
Yes	8	8
No	19	19
4) Did the obstetrician convey information about survival rate in CCAM patients?		
Yes	10	10
No	17	17
5) Which of the following risk were presented to you when communicating diagnosis?		
Intrauterine death	16	16
Death at birth	12	12
Fetal distress	27	27
Polidramnios	0	0
Other	0	0
6) Did the obstetrician recommend pregnancy termination?		
Yes	13	13
No	14	14
7) Who suggested seeking surgical advice?		
Obstetrician	19	19
Hospital web-site	8	8
Other	0	0
8) Which was your major concern since diagnosis?		
Loosing the baby during pregnancy	15	16
Loosing the baby at birth	10	3
Increase of cystic size	2	8
Other	0	0
9)Which was the main emotion you experienced at diagnosis?		
Fear	8	3
Anger	0	4
Sense of guilty	1	0
Disorientation	18	20
Discouragement	0	0
Hope	0	0
Other	0	0
10) Which of the following was the most distressing moment for you?		
Diagnosis	14	18
Time between diagnosis and surgical consultation	12	7
First consultation with the surgeon	0	0
Time after diagnosis	0	0
Follow-up controls	0	0
Delivery	1	2
Other	0	0

Discussion

The majority of prenatally diagnosed CCAM, will have good outcomes, in particular spontaneous in utero regression is now recognized as a common event occurring in up to 43% to 86% of lesions (8). Natural history associated with larger CCAM may be more variable but prognosis remains quite favourable (8, 9). Our series is quite consistent with these findings: as a matter of fact, we did not observe in utero deaths and only two babies underwent surgery at birth.
To our knowledge, few studies (10, 11) have drawn the attention to parents'subjective experience of first communication of diagnosis when a CCAM is discovered in their fetus.
Despite prenatal consultation with the surgeon is nowadays offered to most of the couples after diagnosis of a congenital malformation, such consultations are not subject to legislation and the procedures used to inform prospective parents are not clearly laid down.
Few studies have drawn the attention to how information given may affect decision about whether or not pursuing pregnancy, therefore, message to be given to prospective parents is far from being clear-cut. Furthermore, there are no guidelines concerning which type of information should be given by the sonologist, how it should be given and which competences are required by medical staff (8-10). For the same reason, little is known about relationship between type of anomaly diagnosed in utero and subsequent consultation.
We noted that most of the couples received the diagnosis of CCAM in the period ranging between 22 to 27 weeks of gestation. As expected, in the vast majority of cases it

was the sonologist who had the difficult task to inform prospective parents about the diagnosis in their fetus (12-16). However even if breaking of bad news is considered an emotive subject for healthcare professionals there are no guidelines concerning which type of information should be given by those performing ultrasound when a CCAM is diagnosed.

Only 10 (37%) couples in our sample reported having received information regarding CCAM survival rate by the sonologist first announcing the diagnosis (question 4) and only 8 reported having received information regarding quality of life after surgery.

According to what perceived by the parents, major risks presented to them by the sonologist were: fetal distress (27/27), intrauterine death (16/279, stillbirth (12/27). Therefore, considering natural history of such anomaly, one can conclude, that in a significant number of cases, risks were either"over expressed" or "overperceived" by the parents.

Such lack of prognostic information may be particularly distressing for parents who are at a gestation age at which the option of pregnancy termination (PT) is still legal according to our Contry's law. As previously mentioned, PT was suggested to 13 couples, that is nearly half of the couples returning to questionnaire.

Of note is the adviced rate of termination of pregnancy that was nearly 50% for this fetal malformation as reported by parents. This is unacceptable considering prognosis of CCAM in utero as well as ex utero.

We might speculate that it is the variable in utero expression and evolution of CCAM that may generate uncertainty in the healthcare professional.

Even though parents might have tendency to focus mostly on negative information, these data stress the importance of implementing communication trainings and supervision services for prenatal healthcare professionals that have the difficult and important task of breaking bad news (17-20). Regarding parents' emotional reactions after diagnosis, 18 mothers (66%) and 20 fathers (74%) reported disorientation whereas fear was present in 8 mothers (30%) and 3 fathers (11%). These data emphasize that following an adverse prenatal diagnosis, clinical collaboration between obstetricians, pediatric surgeons and mental health specialists might be the most helpful way to approach couples'needs. Moreover, since the two most stressful moments for parents were communication of diagnosis and time between diagnosis and surgical consultation, it is likely that prompt surgical referral and written material to be given to the couples may be of great help to bridge the gap between diagnosis and consultation with the surgeon.

When analyzing major concerns of parents since diagnosis, 17 mothers and 22 fathers reported fear to lose the baby throughout pregnancy and suggests that further studies are needed to assess how prenatal counselling may affect parents-baby relationship (21-25) since pregnancy. This study has several limitations, **the first and most important being that the questionnaire was not validated; s**econdly, healthcare professionals were not taken into consideration so that only parents'perspective was considered. However, we were able to analyze data from 27 questionnaire which, given the rarity of the disease, represent a significant number of cases. These data provide recommendations for effective prenatal counselling in the setting of a known congenital anomaly from parents's perspective. Whether or not a similar experience can be found in different countries with different practices and, **perhaps, with different congenital anomalies should** deserve further investigation.

Conclusions

Despite course of CCAM diagnosed in usually benign, a significant number of parents appears to receive incomplete information with risk of fetal distress and demise prevailing over survival rate and quality of life. Counselling in this particular setting should be provided only by a multidisciplinary team able to allow the most exhaustive comprehension of the fetal anomaly as well as to offer high-quality supportive care for prospective parents.

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