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Congenital cystic adenomatoid malformation – diagnostic and therapeutic procedure: 8-year experience of one medical centre

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Abstract

Introduction: Congenital cystic adenomatoid malformation (CCAM) is a rare anomaly. The mechanisms and the time at which the abnormality develops are still unclear. The malformation is characterized by the presence of single large or multiple but smaller cysts.

Aim: To present the experience of our medical centre, the Polish Mother's Memorial Hospital – Research Institute.

Material and methods: We analysed the medical records of 32 neonates hospitalized in 2008–2017 at the Department of Paediatric Surgery and Urology ICZMP due to pre- or postnatally diagnosed congenital cystic adenomatoid malformation.

Results: In 2008–2017 32 children were hospitalized at our department due to prenatally diagnosed CCAM. An intrauterine procedure of thoracoamniotic shunting was performed in 13 fetuses. All newborns had chest computed tomography. Surgery had been performed in 21 children. Histopathological analysis of specimens prepared from the resected lung tissue confirmed type 1 CCAM in 4 cases, type 2 CCAM in 11 cases and type 3 CCAM in 1 case. Four patients were diagnosed with a bronchogenic cyst and one with extralobar pulmonary sequestration.

Conclusions: Postnatal management of CCAM depends on the clinical status of the newborn. Newborns without clinical symptoms require radiological diagnostics to confirm the presence of malformation. Surgical treatment is recommended before the age of 10 months. Resection of the malformed lung fragment in patients with CCAM is the first-line treatment. It should be emphasized that despite the tremendous advances in diagnostic imaging, the final diagnosis of CCAM is made based on histopathological findings.

Key words: congenital cystic adenomatoid malformation, thoracoamniotic shunting.

Streszczenie

Wstęp: Wrodzona malformacja gruczołowo-torbielowata płuc (CCAM) jest rzadką anomalią. Obecnie nieznane są zarówno mechanizmy, jak i czas powstania wady. Malformacja charakteryzuje się obecnością pojedynczych, dużych lub bardzo licznych, ale mniejszych torbieli, zwykle obejmujących pojedynczy płat płuca.

Cel: Przedstawienie doświadczenia naszego ośrodka – Instytutu Centrum Zdrowia Matki Polki (ICZMP) w Łodzi.

Materiał i metody: Przeprowadzono analizę dokumentacji medycznej noworodków hospitalizowanych w latach 2008–2017 w Klinice Chirurgii i Urologii Dziecięcej ICZMP w Łodzi z powodu pre- lub postnatalnie rozpoznanej CCAM.

Wyniki: W latach 2008–2017 w klinice hospitalizowano 32 dzieci z powodu prenatalnie rozpoznanej torbielowatości płuc. U 13 płodów wykonano zabieg wewnątrzmaciczny polegający na założeniu shuntu opłucnowo-owodniowego. U wszystkich noworodków wykonano badanie tomografii komputerowej klatki piersiowej. W chwili powstawania pracy zabieg chirurgiczny przeprowadzono u 21 dzieci. Wyniki badań histopatologicznych preparatów wyciętej tkanki płucnej potwierdziły obecność CCAM typu I w 4 przypadkach, typu II w 11 przypadkach, typu III w 1 przypadku. U 4 pacjentów wynik w badaniu histopatologicznym wykazał obecność torbieli bronchogennej, a u 1 – sekwestru zewnątrzpłucnego.

Wnioski: Postępowanie postnatalne zależy od stanu klinicznego noworodka. Noworodki bez objawów klinicznych wymagają diagnostyki radiologicznej w celu potwierdzenia obecności wady. Przeprowadzenie zabiegu rekomendowane jest w 10. miesiącu życia. Wycięcie fragmentu zmienionego płuca w przebiegu CCAM jest postępowaniem z wyboru. Należy podkreślić, że pomimo ogromnego postępu w diagnostyce obrazowej ostateczne rozpoznanie CCAM ustala się na podstawie badania histopatologicznego.

Słowa kluczowe: wrodzona malformacja gruczołowo-torbielowata płuc, shunt opłucnowo-owodniowy.

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Introduction

Congenital cystic adenomatoid malformation (CCAM) is a rare anomaly with reported incidence ranging from 1 in 25,000 to 1 in 35,000 live births [1]. It is usually an isolated disorder, although its coexistence with other lung defects, e.g. pulmonary sequestration, and cardiac or renal defects, has been reported [2]. The mechanisms and the time at which the abnormality develops are still unclear. Most likely, this defect is a result of arrested lung development in the early phase of fetal life (6–15 gestational weeks (GW)), which results in hypertrophy of bronchia/bronchioles and mesenchymal tissue and failure to develop alveoli [3]. Malformation is characterized by the presence of single large or multiple but smaller cysts usually involving a single lobe of lung. Multilobar lesions involving both lungs are very rare. A tendency for spontaneous resolution of the microcystic types of CCAM (usually after 28 GW) has been reported [4]. However, the postnatal regression of lesions is unlikely. The macrocystic type of CCAM does not tend to regress, and large fluid-filled cysts may compress the healthy lung parenchyma, causing its hypoplasia or mediastinal shift and compression of the lower vena cava, contributing to fetal cardiovascular insufficiency and to the development of generalised fetal hydrops [5]. Hydrops is associated with the risk of fetal death. If there is a risk of such complications, antenatal therapy should be considered [5].

In the postnatal period the key clinical symptoms of CCAM in the newborn depend on the type and severity of lung lesions. Pathological cysts filled with air that are in contact with normally developed bronchia and bronchioles can rapidly compress the remaining lung parenchyma, cause mediastinal shift, provoke disorders of venous circulation (venous flow), and lead to respiratory/cardiovascular insufficiency. After the neonatal period abnormal lung tissue may become a reservoir of secretions accumulated in cystic spaces and a source of recurrent infections [2]. Cases of malignant transformation in the abnormal lung tissue have also been reported [6].

In 1977 an American pathomorphologist, JT Stocker, described 3 histopathological types of CCAM. In 2002 he added another two types to the list (0 and 4), and his classification system is used to this day [7] (Table I). The most common types of CCAM are macrocystic type 1 (60%) and microcystic type 2 (15–30%) [7] (Figs. 1–3). Treatment for CCAM involves the resection of malformed lung tissue.

Table I. Stocker histopathological classification of CCAM (2002)

Type 0	Development arrested at the stage of trachea/bronchia formation; tracheal epithelium. Cysts smaller than 0.5 cm; tracheal epithelium, presence of cartilage
Type 1	Development arrested at the stage of bronchia formation; bronchial epithelium. Large 4–10 cm cysts, cartilage rarely present, squamous-like epithelium
Type 2	Development of the bronchial tree arrested at the glandular stage. Multiple cysts < 2.5 cm, covered with columnar epithelium
Type 3	Development of the bronchial tree arrested at the glandular stage, typical adenomatoid malformation. Multiple cysts < 1.5 cm, covered with columnar epithelium
Type 4	Development arrested at the stage of bronchia formation; acinar epithelium. Cysts size 2–4 cm (pleuropulmonary blastoma – PPB), cartilage absent

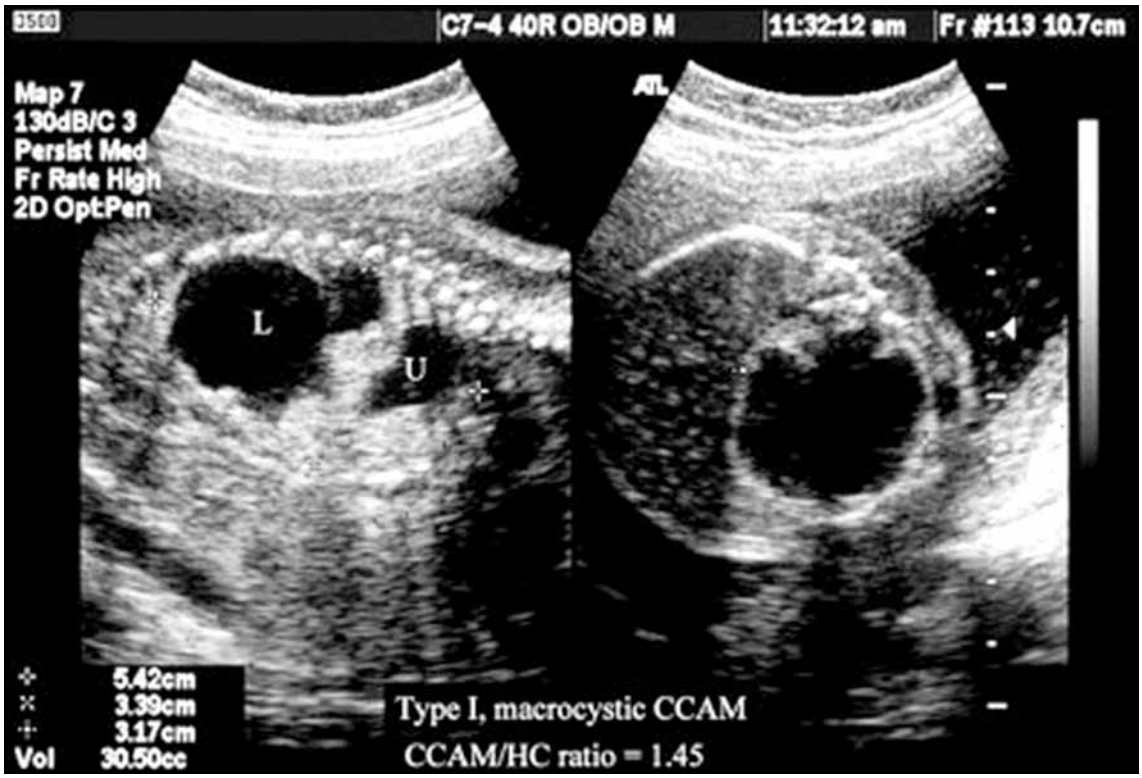


Fig. 1. Macrocystic CCAM (USG scan)

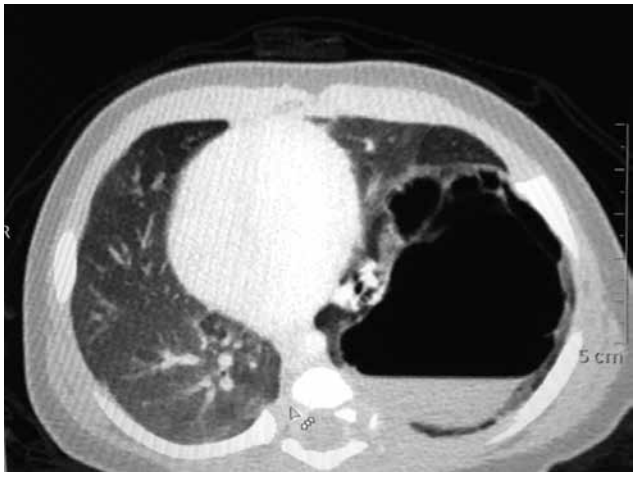
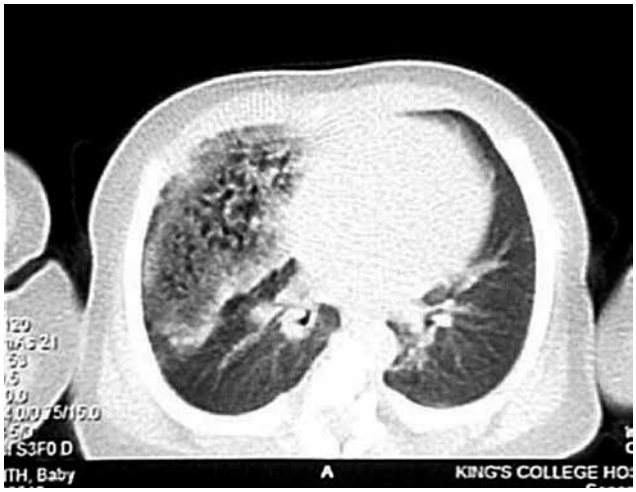


Fig. 2. Macrocystic CCAM (CT scan)



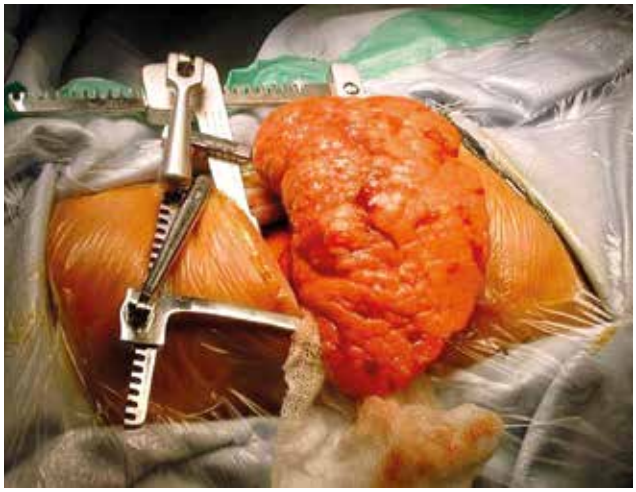


Fig. 3. Microcystic CCAM (CT scan and intraoperative imaging)

Aim

The aim of this study is to present the experience of our medical centre, the Polish Mother's Memorial Hospital – Research Institute in Lodz (ICZMP), in the pre- and postnatal diagnosis and treatment of newborns and infants with CCAM.

Material and methods

We analysed the medical records of neonates hospitalized in 2008–2017 at the Department of Paediatric Surgery and Urology ICZMP in Lodz due to pre- or postnatally diagnosed congenital cystic adenomatoid malformation. The collected data concerned:

– The prenatal period, including the gestational age at the time of diagnosis of malformation, the type of malformation based on fetal ultrasonography, and indications, type and duration of prenatal intervention.
– The postnatal period: gestational age, body weight, Apgar score at birth, sex of the child, clinical assessment in the first days of life, type and location of the malformation in the computed tomography (CT) scan, presence of accompanying defects, age of the patient at the time of surgery, findings from histopathological examination of the resected fragment of the lung, and existing accompanying defects. Type of malformation was assessed using the Stocker classification system (2002) covering five types of CCAM (0–4).

Results

In 2008–2017, 32 children (17 girls, 53%; 15 boys, 47%) were hospitalized at the Department of Paediatric Surgery and Urology ICZMP in Lodz due to prenatally diagnosed congenital cystic adenomatoid malformation. Malformation was diagnosed and monitored using ultrasound scan (US) (Fig. 1 – fetal US). Gestational age of the fetuses at diagnosis was in the range of 20–33 GW (mean 26 GW). Echocardiography was also performed in 23 (72%) fetuses. Examination revealed heart defects in 4 children: cardiomyopathy (n = 1), ventricular septal defect (n = 2), and aortic stenosis with dextrocardia (n = 1). An intrauterine procedure of thoracoamniotic shunting was performed in 13 fetuses. The procedure was indicated due to an expanding cystic lesion that compressed the residual lung parenchyma and caused mediastinal shift. In addition mothers were diagnosed with progressing polyhydramnios.

At birth the gestational age of the paediatric patients ranged between 30 and 40 GW (mean 38 GW). Most children from the study group were born full-term (n = 26). In 6 cases, there was an indication for the early termination of pregnancy. This group included 3 newborns who in the prenatal period underwent the procedure of thoracoamniotic shunt placement. Indications for the early termination of pregnancy included threatening fetal asphyxia (n = 1), cardiovascular disease of the mother (n = 1), orthopaedic problems of the mother (n = 1), and spontaneous onset of preterm labour (n = 3). Caesarean section was performed in 20 cases and 12 children were delivered vaginally.

The general health of most newborns from the study group (n = 25) after delivery was good (Apgar score 9–10 points). This group included 9 newborns who had a prenatal procedure of thoracoamniotic shunt placement. Six newborns, including 2 treated prenatally, received moderate scores due to respiratory effort and suspected congenital infection (Apgar 7–8 points). Serious or very serious

health problems with severe respiratory insufficiency and the need for intubation were diagnosed in one neonate (Apgar 1–6) born with asphyxia at 30 weeks of gestation. In this newborn, an intrauterine procedure was performed, complicated by bleeding into the pleural cavity.

All newborns had chest CT, usually on day 7 after birth (Fig. 2). Depending on the cyst size newborns were diagnosed with macrocystic CCAM (n = 13) or microcystic CCAM (n = 11). In other children (n = 8) CT scan revealed solid cystic lesions.

Postnatal diagnostic procedures also showed the presence of coexisting defects in 8 (25%) children with CCAM, including heart defects (n = 4), cleft palate (n = 1), CNS cysts (n = 1), urinary tract defects – PCS dilatation (n = 1), and one child was diagnosed with haemophilia. Of the above, only heart defects were diagnosed prenatally.

By the time of manuscript preparation, surgery had been performed in 21 children. In 6 newborns the procedure was carried out between 8 and 20 days of life due to progressing respiratory insufficiency. Other patients (n = 15) were qualified for elective surgery between 2 and 12 months of life. Non-operated patients (n = 11) are under the care of other medical centres and/or await surgical treatment. In 16 patients the procedure was performed by thoracotomy and in 5 by thoracoscopy. In 1 case the operator had to proceed from thoracoscopy to thoracotomy. During the surgery lobectomy was performed in 14 children and segmentectomy in 6 children. In 1 patient extrapulmonary sequestration was detected during thoracoscopy and the malformation was completely resected. Postoperative pleural drainage was maintained in all patients for 1 to 5 days (mean: 3 days). All children received perioperative and postoperative antibiotic therapy for 5 days on average (range: 3–7 days). The postoperative period was uneventful in 17 (81%) children. During the post-operative period one child suffered a cardiac arrest with leakage of lymphatic fluid into the chest cavity. Despite preventive antibiotic therapy postoperative deterioration of respiratory function due to systemic infection was observed in 3 children. One of these children was preoperatively diagnosed with pulmonary infection. No deaths in the study group were reported.

Histopathological analysis of specimens prepared from the resected lung tissue confirmed type 1 CCAM in 4 cases, type 2 CCAM in 11 cases and type 3 CCAM in 1 case. Based on histopathological examination 4 patients were diagnosed with a bronchogenic cyst and 1 with extralobar pulmonary sequestration (Table II).

Discussion

With advances in fetal ultrasonography and recently introduced fetal magnetic resonance imaging, most congenital lung defects can be diagnosed in the early intrauterine period. It should be emphasized that lesions detected in the fetal ultrasonographic image of lungs are not specific just for CCAM. Increased echogenicity of the lung and the presence of cysts are described in other disorders, e.g. pulmonary sequestration, bronchogenic or enterogenous cysts, and in congenital lobar overinflation [8]. Therefore, a more generalized term such as congenital lung malformation (CLM) is used increasingly often. Imaging studies, including ultrasonography, CT, and magnetic resonance (MR), are used to distinguish macrocystic, microcystic or a mixed, solid cystic type of malformation [9].

According to reports, fetal lung defects are usually diagnosed between 18 and 34 GW [8]. In our study group CCAM was diagnosed on average at 26 GW. The evolution of the malformation is monitored in subsequent weeks of pregnancy, not only for prognostic purposes, but also for proper planning of fetal therapy. In the study group, a prenatal intervention involving shunt placement between the lumen of the lung cyst and amniotic fluid was performed in 13 fetuses. Indications for surgery included expansively growing cyst compressing the residual lung parenchyma, mediastinal shift in the fetus, and progressive hydramnios in the mother. Echocardiography was performed before surgery in all fetuses to confirm their cardiac status. This protocol is consistent with the latest recommendations [10]. Reports from leading medical centres offering fetal therapy in different countries have indicated that about 14% of fetuses with CCAM require prenatal intervention, most commonly thoracoamniotic shunting, which significantly improves the survival of fetuses with CCAM, and/or the possibility of steroid therapy [10]. For several years, attempts have also been made to perform radical intrauterine operations – removing pathological lung tissue in CCAM fetuses, particularly those at risk of developing or with existing generalized hydrops [10]. There are many questions as to the possibility of spontaneous regression of lung lesions in the prenatal period. Recent studies show that microcystic lesions previously described as lesions tending to resolution are often secondary to bronchial obstruction and fluid accumulation, rather than actual CCAM lesions [6].

Data on age and body weight of newborns at birth in the study group were consistent with those reported by other authors [11]. The gestational age of paediatric patients ranged between 30 and 40 GW (mean 38 GW), and the mean birth weight was 3450 g (2730–3800 g). In 6 cases there were indications for the early termination of pregnancy due to threatening fetal asphyxia or the medical condition of the mother. This group included 3 newborns who received fetal therapy (Table III). Although some reports show that a fetal lung defect is not an indication for Caesarean section [12], most children (n = 20) from the study group were born in this way.

In neonates, the presence and severity of respiratory distress syndrome due to the extent and severity of lung lesions is crucial [13]. The rapid enlargement of pathological cystic spaces as a result of their aeration, compression of the normally developed lung parenchyma, and mediastinal shift are the most important factors worsening the general health status and increasing the severity of symptoms of respiratory insufficiency in newborns with CCAM. Pulmonary hypoplasia and the development of pulmonary hypertension or the coexisting defects may further aggravate the patient's condition [10]. According to some reports,

Table II. Data of radiological and histopathological diagnosis

Ordinal	Type of CCAM in prenatal USG	Localisation in USG	Type of CCAM in CT	Localisation in CT	Time of operation	Histopathology
1	Macrocystic	Left lung	Macrocystic	Left lung, lower lobe	20 days	II
2	Microcystic	Left lung, upper lobe	Microcystic	Left lung, upper lobe	5 months	II
3	CCAM/ sequestration	Left lung	Microcystic	Right lobe, segment 10	Lack of contact	Lack of contact
4	No result	No result	Macrocystic	Left lung, lower lobe	Under the care of another department	–
5	Microcystic	Right lung, middle lobe	Microcystic	Right lung, segment 10	8 days	II
6	No result	No result	Microcystic	Left lung, upper lobe	13 days	II
7	No result	No result	Microcystic	Right lung, upper lobe	4 months	II
8	Microcystic	Left lung	Microcystic	Left lung, segment 9 and 10	10 months	II
9	No result	No result	Macrocystic	Right lung, segment 8, 9, 10	2 months	Bronchogenic cyst
10	Macrocystic	Right lung	Solid cystic lesions	Right lung	2 months	II
11	No result	No result	Microcystic	Left lung, segment 6	Lack of contact	Lack of contact
12	Macrocystic	Left lung, upper lobe	Macrocystic	Left lung, segment 1 and 2	Under the care of another department	Under the care of another department
13	Macrocystic	Right lung	Solid cystic lesions	Right lung, segment 4 and 5	Lack of contact	Lack of contact
14	Microcystic	Right lung	Macrocystic	Right lung, upper and middle lobe	2 months	I
15	Microcystic	Left lung	Macrocystic	Left lung, upper lobe	4 months	I
16	Macrocystic	Right lung	Macrocystic	Right lung, segment 6 and 10	Under the care of another department	Under the care of another department
17	No result	No result	Microcystic	Right lung, segment 6, 9 and 10	4 months	II
18	No result	No result	Solid cystic lesions	Left lung, segment 1/2 i 3	No result	Passed to another department due to haemophilia
19	No result	No result	Macrocystic	Right lung, segment 2	7 months	Bronchogenic cyst
20	No result	No result	Solid cystic lesions	Right lung, segment 9 and 10	4 months	Pulmonary sequestration
21	Macrocystic	Left lung, middle part	Macrocystic	Left lung, lower lobe	Further diagnostics	Waiting
22	Macrocystic	Left lung, upper lobe	Macrocystic	Left lung, lower lobe	8 days	I
23	No result	No result	Macrocystic	Right lung, segment 2 and 6	3 months	II
24	No result	No result	Microcystic	Right lung, segment 6, 8, 9 and 10	Under the care of another department	Under the care of another department
25	Microcystic	Right lung	Macrocystic	Right lung, upper and lower lobe	16 days	I
26	No result	No result	Solid cystic lesions	Left lung, segment 9 and 10	4 months	Bronchogenic cyst
27	Microcystic	Right lung	Microcystic	Right lung, segment 6, 9, 10	5 months	III
28	No result	No result	Solid cystic lesions	Left lung, upper lobe	10 months	Bronchogenic cyst
29	No result	No result	Solid cystic lesions	Right lung, lower lobe	12 months	II
30	Microcystic	Left lung	Solid cystic lesions	Left lung, segment 1, 2, 3	10 days	II
31	Sequestration	Right lung	Microcystic	Right lung, segment 10	Classified	Classified
32	Microcystic	Left lung	Macrocystic	Left lung, segment 10	Classified	Classified

Table III. Data of prenatal care

Ordinal	Gender	Gestational age at diagnosis	Intrauterine procedure	Gestational age of procedure	Coexisting defects	Gestational age	Indication for the early termination	Apgar (1/5 minute)
1	F	29	–	–	–	39	–	9
2	M	23	–	–	CoA	37	Spontaneous onset of preterm labour	8/8
3	F	26	–	–	–	40	–	9/9
4	F	26	–	–	Heart hypertrophy	38	–	9/9
5	F	24	Yes	24	–	35	Orthopaedic problems of the mother	7/7
6	F	23	Yes	23	–	38	–	9
7	M	–	–	–	–	39	–	9/9
8	F	21	–	–	–	39	–	9
9	F	–	–	–	–	40	–	8
10	F	32	Yes	32	–	39	–	8/8
11	F	25	–	–	–	37	Cardiovascular disease of the mother	9/9
12	M	28	Yes	28, 37	–	40	–	9/10
13	F	20	Yes	22	Cleft palate	40	–	10
14	M	31	Yes	31 and 32	Brain cyst	40	–	9
15	F	33	Yes	33	–	38	–	9
16	M	23	Yes	23	–	38	–	8/9
17	M	b.d.	–	–	–	39	–	8/9
18	M	25	–	–	Haemophilia	37	Spontaneous onset of preterm labour	8/8
19	F	28	–	–	–	39	–	10/10
20	M	21	–	–	VSD	39	–	8/8
21	M	22	Yes	26, 27	–	30	Threatening fetal asphyxia	1/1
22	M	29	Yes	32	–	38	–	9
23	F	29	–	–	–	38	–	10
24	F	21	–	–	–	40	–	9
25	M	25	Yes	25	–	37	Spontaneous onset of preterm labour	9
26	M	b.d.	–	–	–	38	–	9
27	F	33	Yes	33	Widening of the pelvic system	38	–	9
28	M	28	–	–	–	38	–	9
29	M	29	–	–	–	39	–	10
30	F	28	Yes	32	VSD	40	–	9/9
31	M	25	–	–	–	40	–	9/9
32	F	24	–	–	–	40	–	9/9

approximately 10–25% of newborns with congenital cystic lung lesions require surgical intervention in the early days of life [11]. Most cases of CCAM in the neonatal and early-infant period are asymptomatic [13]. The general health of most newborns in the study group (n = 25) after delivery was good (Apgar score 9–10 points). The health of 6 newborns was scored as moderate (Apgar 7–8 points) due to respiratory effort and suspected congenital infection. Serious or very serious health problems (Apgar 1–6) with severe respiratory insufficiency and the need for intubation were diagnosed in one newborn who was treated in utero. At present, most authors indicate that bilateral lung lesions and the development of non-immune hydrops are very poor prognostic features and are associated with high mortality, especially in fetuses that received no prenatal intervention [14]. The extent and type of lung lesions, mediastinal shift

and hydramnios may be considered negative prognostic factors, although many of the affected fetuses do not have clinical symptoms of malformation [15]. In the study group 13 newborns who underwent intrauterine therapy required surgery in the first 3 weeks of life due to progressing symptoms of respiratory insufficiency. Other newborns with asymptomatic malformation in the neonatal period received elective surgical treatment.

Due to the evolution of the image of lung lesions observed especially in the last weeks of pregnancy, postnatal computed tomography of the lung is recommended in all newborns with this anomaly [15]. Chest X-ray, especially if done in the first few days of life and in patients with small, limited lung lesions, may not visualise them. Magnetic resonance is a more useful test for the prenatal diagnosis of lung defects [16]. The time of CT examination mainly depends on the clinical status of the newborn. In neonates with clinical symptoms such as respiratory disorders, CT is performed in the first days of life, and in asymptomatic patients usually in the first month of life. In our study group all newborns had chest CT, usually on days 6–7 of life. This study confirmed the prenatal diagnosis of cystic lung lesions in all examined neonates, and the CT image in most cases (n = 19) was consistent with the ultrasound image acquired prenatally. Based on CT imaging macrocystic CCAM was confirmed in 13 children and microcystic CCAM in 11 children. In other patients (n = 8) the CT scan revealed solid cystic lesions. Reports by other authors and the experience of our medical centre show that in some cases certain lung lesions are difficult to differentiate, even if CT is employed. This especially concerns the segmental areas that may suggest small airway obstruction, lobular emphysema, or CCAM [2]. In many cases resected lung fragments in histopathological examination turned out to be lesions associated with emphysema or bronchogenic cysts. Hybrid lesions are reported increasingly more often (about 25% cases), and they contain features of adenoid cystic lesions and pulmonary sequestration, or segmental bronchial obstruction with or without emphysema and CCAM [17].

Lung lesions in CCAM usually affect a single lobe, usually the lower one and more often the left lung. Bilateral pathology is very rare [18]. In the study group lung lesions were usually located in the right lung (n = 17), mainly in segments 6, 9, 10 of the lower lobe (n = 10), and less frequently in the upper lobe. In 15 children lesions were detected in the left lung, usually in the lower segments of the lower lobe. In the study group we did not observe prenatal or postnatal regression of pulmonary lesions, although cases of prenatal resolution are reported in the literature [19].

Children diagnosed with CCAM often have additional anomalies. In our study group most of these were heart defects, usually correlated with the solid type 2 CCAM. The coexistence of various defects suggests the same pathomechanisms underlying their abnormal embryogenesis [20].

Recent advances in thoracic surgery and anaesthesia have minimized the mortality of newborns and infants with congenital cystic pulmonary lesions. Today it is clear that pathological lung tissue has to be removed, and the only controversy is about the time of the procedure and its scope. In neonates with clinical symptoms the surgery should be performed after previous diagnostic imaging and stabilization of vital parameters [15]. In the study group 6 children were operated on in the early days of life due to progressing symptoms of respiratory insufficiency. Surgery was performed between 8 and 20 days of life. Other children (n = 15) were operated on between 1 and 12 months of life (mean: 5 and 6 months). Two children who did not show clinical symptoms of malformation during the monitoring before surgery were treated for lung infection. These children were operated on at the age of 2 or 4 months after the resolution of the lung infection. In the early postoperative period one of these patients developed respiratory insufficiency. A meta-analysis carried out in 2009 by Stanton *et al.* revealed that the age of 10 months and younger is the optimal time for the surgical treatment of children with asymptomatic cystic malformation of the lung [21]. During this time most patients begin to show the first clinical symptoms of the malformation, most often in the form of pulmonary infections. Many surgeons recommend an elective procedure at the age of 6 months or earlier, before the onset of pulmonary complications [15]. Many studies suggest that patients with CCAM-related clinical symptoms, especially pulmonary infections, have a significantly higher incidence of surgical and postoperative complications compared to patients with asymptomatic defects [22]. There are also reports indicating that the early resection of abnormal lung tissue allows for better development of the unaffected lung [22]. Only a small proportion of children with a minor segmental lesion may be monitored for a longer time [16]. If the treatment is postponed, the parents of the patient should be informed and made aware of the adverse consequences of the disease, primarily the increased risk of pulmonary infection or malignant transformation.

Many surgeons believe that in cases where pulmonary lesions are limited to a single lobe, the most appropriate surgical procedure is to remove the lesion completely (finding a clear border between CCAM and normal lung parenchyma may be difficult in macroscopic examination) and reduce the risk of early pleural effusion often observed after segmentectomy [23]. Thoracoscopic minimally invasive techniques are applied increasingly more often, both for lobectomy and segmentectomy, and are especially recommended in children with asymptomatic CCAM. The extent of pulmonary lesions and, above all, respiratory disorders in neonates are factors limiting the possibility of performing thoracoscopic procedures. A history of pulmonary infections significantly increases the risk of intraoperative conversion to classic thoracotomy and the possibility of postoperative complications including pneumothorax, bronchial fistula and septicaemia [24]. Children who have undergone segmentectomy (n = 4) in our centre are under constant ambulatory care, and control CT studies have not revealed pathological lesions in the surgically treated lung.

No deaths were reported in the study group. According to different authors, a significant improvement in the mortality rate has been seen over the past few years in comparison with previous studies [12]. The deaths of children with CCAM are usually linked to postoperative complications. The size of lesions is considered an important prognostic factor for survival [16].

According to the literature, type 1 CCAM is most commonly diagnosed in the histopathological examination [17]. In our study, however, Stocker's type 2 CCAM (2002) was diagnosed most frequently. Baird *et al.* point to the fact that the image of cystic lung lesions in pre- and postnatal studies often does not correspond to the lesions found later in histopathological studies. Some reports suggest that the discrepancy rate for diagnostic findings is up to 17% [8]. This may be due to the unclear border between macroscopic and microscopic images, as well as the lack of a clear border in the histopathological image of particular CCAM types. Hybrid lesions that contain features of CCAM with sequestration are being reported with increasing frequency [17]. Diagnostic imaging studies often detect lesions in various respiratory phases of the lung and varying degrees of pathological cyst filling [25]. In the group of surgically treated children an initial diagnosis of CCAM made based on imaging studies (CT) was confirmed in most cases (n = 16) after histopathological examination. In four cases, histopathological examination revealed the presence of intrapulmonary bronchogenic cyst, a rare congenital malformation resulting from abnormal development of the bronchial tree, resembling type 1 CCAM in the CT image. In one case the solid cystic lesion detected in the CT image was in fact associated with pulmonary sequestration.

Disclosure

Authors report no conflict of interest.

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