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Radiological diagnostics in neonates with different types of congenital cystic adenomatoid malformation of the lungs (CCAM) treated in Polish Mother's Memorial Hospital Research Institute (PMMHRI) in 1991-2005

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Summary

Background: Congenital cystic adenomatoid malformation of the lungs (CCAM) is a rare congenital malformation of the respiratory tract. Authors present possibilities of the diagnostics of neonates with presumed CCAM based on radiological and morphological assessment of the respiratory tract. Clinical course of the CCAM may vary from uneventful to serious with different stage of the respiratory distress.

The aim of the study was to establish diagnostic and clinical criteria of CCAM for neonates based on postnatal diagnostics in the reference centre.

Material/Methods: We studied 27 cases of neonates with different types of CCAM (type I, II, III) who had been diagnosed and/or treated in the Neonatal Department of PMMHRI in 01.01.1991-31.03.2005. The diagnosis was established based on clinical course of the malformation, chest x-ray and CT of the neonate's lungs, autopsy and/or histopathology.

Results: In the Neonatal Department of PMMHRI in 01.01.1991-31.03.2005 we observed 8 cases of CCAM type I (29,7%), 10 cases of CCAM type II (37%) and 9 cases of CCAM type III (33,3%). Diagnostic criteria were established based on radiological diagnostics of 17 cases and were confirmed by pathology. In 10 remaining cases of CCAM diagnosis was established by authopsy. Differential diagnosis included diaphragmatic hernia, bronchogenic cyst, enterogenic cyst, lung sequestrqtion, congenital lobar emphysema, hypoplasia or agenesis of the lungs, pneumonia/RDS. The guideline was established.

Conclusions: 1. In case of CCAM suspicion monitoring in reference center is required 2. Surgical treatment should be applied based on the postnatal radiological diagnosis. 3. In case of CCAM in neonate complete differential diagnosis is required. 4. Asymptomatic course of CCAM or with a very few signs from respiratory tract can be difficult for identification both clinical and radiological.

Key words: congenital cystic adenomatoid malformation of the lungs (CCAM) • imaging radiological diagnostics • neonate • guideline

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Background

Congenital cystic adenomatoid malformation (CCAM), in latin – *dysplasic pulmonis cystica*, is an infrequent congenital malformation of the respiratory tract, which can be diagnosed correctly in the neonatal period [1, 2, 3, 4, 5, 6, 7, 8].

It was first described in 1949 by Chin and Tang [9] based on anatomopathological examination performed in a neonate with generalized swelling and one lobe of the lung cystically changed.

In 1977 Stocker [10, 11] et al., based on histopathology images, distinguished 3 types of CCAM, which differ in size of the cyst, clinical course and prognostic indicators.

CCAM type I: Massive multiple cysts of diameter > 3 cm to 10 cm or a single cyst surrounded by smaller cysts which can fill up the lung. This form has the best prognosis and is the most common radiological type in children.

CCAM type II: small multiple cysts with diameter of < 3 cm. Survival in this case depends on associated defects (around 56%) and is a rarely considered radiological type.

CCAM type III: solid lesions, often bilateral, with diameter of < 0,5cm. It is the rarest radiological type in children and has poor prognosis.

Suspicion of CCAM in the neonatal period requires monitoring at a reference centre. RTG examination of thorax is performed in the postnatal period, although the intravital confirmation is in changes visible in CT image of the lungs [7, 12, 13, 14, 15].

The clinical course of CCAM can vary from asymptomatic to different levels of respiratory malfunctions. Treatment consists in resection of the changed lobe. Final survival depends on the degree of pulmonary underdevelopment and the level of maturity of the lungs [1, 2, 4, 7, 16, 17, 18, 19].

Aim of the study

Setting the diagnostic and clinical criteria for neonates with various forms of CCAM based on the postnatal diagnostics at the Polish Mother's Memorial Hospital Research Institute (PMMHRI).

Materials and methods

The studied clinical material consisted of 27 cases of different forms of CCAM (type I, II, III according to Stocker), diagnosed and/or treated at The Neonatology Department of PMMHRI between 01.01.1991 and 31.03.2005.

In some cases the lung anomaly was suspected prenatally. The diagnosis was made or verified based on:

- clinical image of the defect
- roentgen examination of neonate's thorax
- computed tomography scan of neonate's lungs
- autopsy and /or histopathology exam

After delivery all neonates underwent transfontanelle and abdominal USG exams, along with ECHO examination of the heart in order to exclude other abnormalities.

The neonates discharged from Neonatology Department of PMMHRI after successful operations on CCAM continued to be under further clinical observation of many specialists of our institute.

Results

27 cases of different types of CCAM were noted at the Neonatology Department of PMMHRI in 1991–2005, including type I – 8 cases (29,7%), type II – 10 cases (37%), type III – 9 cases (33,3%).

Diagnostic criteria for CCAM type I were established based on radiological imaging diagnostics (RTG+CT) proceeded in eight neonates.

In roentgen imaging examinations of thorax, unilateral cystic changes, in most cases – left sided, were stated in the lungs of all patients.

In 5/8 of the cases massive aerocele (max diameter of 40x70 mm), and in 3/8 – multiple cysts of different sizes with maximum diameter of over 3 cm, were described. The depicted changes moved the heart and mediastinum to the opposite side in most cases (fig. 1).

In two cases the result of roentgen examination was not unequivocal. In one case the radiologist suggested differentiating the cysts from diaphragmal hernia, in the second he described the presence of cyst and suspected pneumonia with pneumothorax.

The diagnosis of CCAM type I was made on the basis of CT scan of the lungs performed in 7 neonates (fig. 2), and RTG of the thorax in 1 patient. Definitive diagnosis was confirmed by histopathology examination in 7/8 of cases, while in 1/8 – by autopsy (fig. 3).

In echocardiographic exams of 8 neonates the structure of heart was normal, but functional abnormalities were observed in 7/8 of patients – one showed functional tricuspid incompetence (IT) and vestigial pulmonary valve incompetence (IP) + FoA, in 3 neonates FoA-type interatrial septal defect and Botall's persisting arterial duct (persistent ductus arteriosus -PDA), in single cases – the FoA (3x) and the features of hyperkinetic circulation with disproportion within the cardiac cavities were stated in one child.

Five neonates demonstrated severe respiratory failure with the necessity of respiratory support and required multidirectional treatment at the intensive neonatal care unit (INCU). One of them died in 17th hour of its life. In other two, the tachypnoe-type respiratory malfunctions were observed, while one of the neonates was hardly symptomatic.

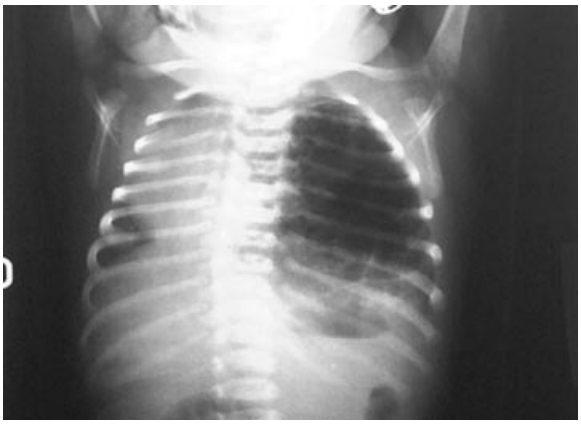

Figure 1. Chest x-ray of the neonate (CCAM type I) – huge cyst in the left side of the chest shifting mediastinum to the right.

Seven neonates from the described group underwent surgical treatment, most (6/7) at the Department of Cardiosurgery of PMMHRI. All operations were performed within the 1st month of their life, on average on the 10th day.

Seven children remained under further clinical observation of our centre, on average for 7 years (min 10 months, max 8 years).

Physical development of children after operations for CCAM type I was assessed as normal. The body mass was between 10 and 75 centile of the norm, while the height was between 25 and 50 centile of the norm according to the criterion of centile charts.

Psychokinetic development of children, evaluated with the use of Denver scale, in 6 cases was adequate to age and only in one case it was retarded (in a child with posthepatitic hydrocephalus, who was treated outside the PMMHRI).

In the medical history, a few children suffered from recurrent urinary tract infections. Two patients required treatment with oral hematinic drugs due to iron deficiency anemia.

Diagnostic criteria for type II of CCAM were set on the basis of radiological imaging (RTG + CT) in 9 neonates, while in one neonate it was based on autopsy.

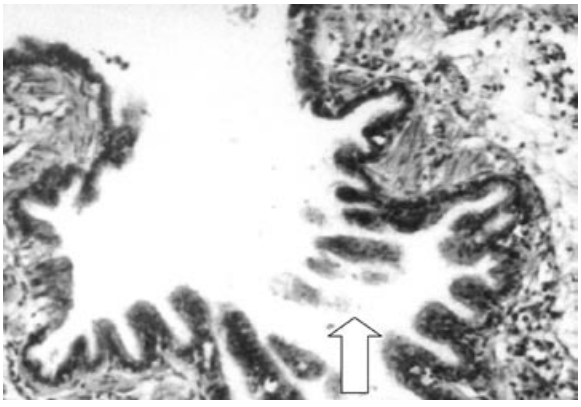

Figure 3. Pathology (CCAM type I) – huge cyst lined with cuboidal epithelium.

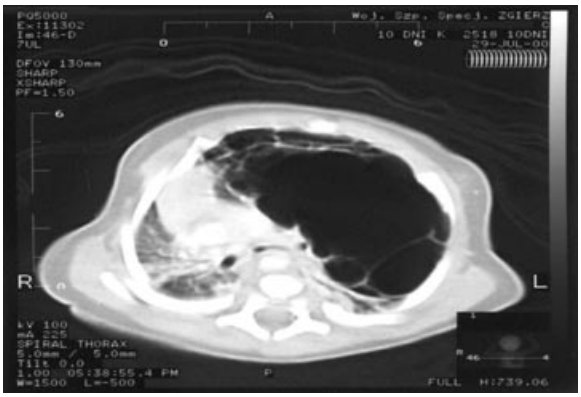

Figure 2. CT of the lungs of the neonate (CCAM type I) – huge cyst in the left side of the chest shifting mediastinum and the heart to the right.

In the roentgen examinations of the thorax of 8 neonates unilateral cystic changes in the lungs were described (8/10). In one child the cystic lesions in the lungs were bilateral.

In 3/10 of cases the roentgen image of the thorax showed multiple unilateral cysts with the diameter of less than 3 cm (fig. 4). In the remaining 6/10 of cases radiological images were not unequivocal. The radiologist suspected pneumonia 4 times (in one case – with emphysematous bulla? pneumothorax?, in the second – with cysts, in the third – nonemphysematic changes, in the next he described airlessness of the superior lobe of left lung with the dislocation of the heart and mediastinum, while in another – he suggested diaphragmal hernia or cysts).

In the neonate with bilateral cystic changes of lungs the differentiation included staphylococcal pneumonia with emphysematous bullas.

The definite diagnosis of CCAM type II was made on the basis of CT of the lungs in 7 neonates (fig. 5), RTG of the thorax in 2 neonates and in one case – the autopsy exam (fig. 6).

The intra-operative histopathology examination in one of the nine operated neonates revealed intralobar sequestration, apart from confirming the CCAM type II.

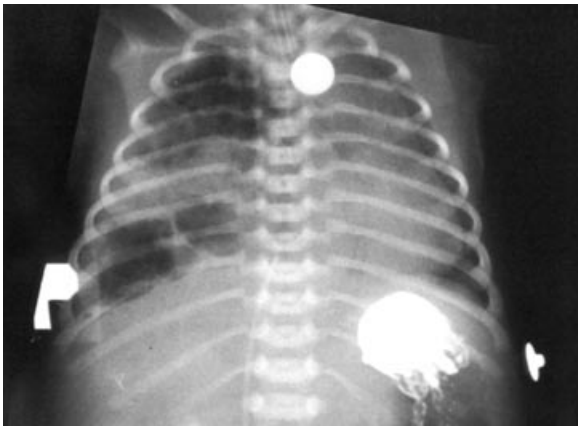

Figure 4. Chest x-ray of neonate (CCAM type II) – multiple cystic changes in the right lung.

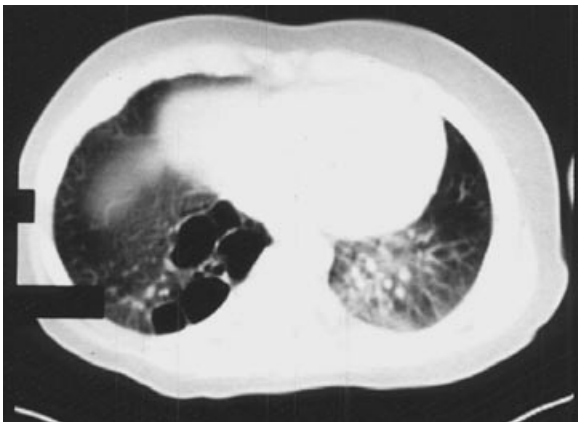

Figure 5. CT of the lung of neonate (CCAM type II) – multiple cystic changes in the right lung.

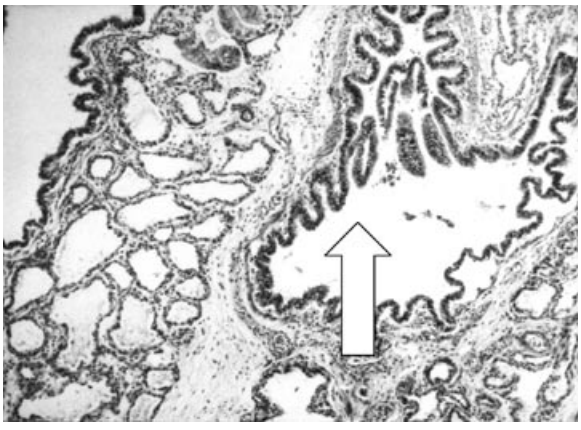

Figure 6. Pathology (CCAM type II) – cyst lined with cuboidal epithelium – arrow.

The echocardiographic examination of nine neonates showed proper heart structure, while in one case there were changes in form of functional tricuspid incompetence (IT) +FoA + PDA, and in single cases – the Botall's persisting arterial duct (PDA) (2 times) and foramen ovale (FoA) – type of interatrial septal defects (3 times).

In 5 neonates the symptoms of acute respiratory failure with necessity for respiratory support at INCU were stated postnatally, while in 3 the tachypnoe type of respiratory failure was observed. In one of the patients the clinical course was asymptomatic, periodically-hardly symptomatic.

In the described group of patients with CCAM type II, there was one death declared in the 2nd hour of life of a premature baby, born in the 29th week of pregnancy, with fetal hydrops, massive ascites, arrhythmia (clinical recognition), bilateral hypoplasia of the lungs (autopsy recognition).

Among the neonates with CCAM type II, 7 underwent operations in the 1st month of life, on average – on the 13th day of life (min 1st day, max 35th day), while the remaining 2 patients – in later time (4th, 12th month of life).

Nine children with CCAM type II remained under further clinical observation for 10 years on average (min 12 months, max 10 years).

The somatic development evaluated according to the criterion of the centile charts, as well as the psychokinetic development evaluated according to the Denver scale was normal in 9 children. A funnel-shaped thorax was stated in four of them.

Infections of superior respiratory tracts recurred in single cases. In one child the symptoms of bronchiopulmonary dysplasia requiring proper treatment were observed, while in the other – nutritional allergy treated with an eliminating diet, and a girl of 4 was diagnosed with bronchial asthma with grass pollen allergy, and currently receives bronchodilators, periodically – the steroids.

Diagnostic criteria for CCAM type III were set on the basis of autopsy examinations in all cases. In 7 cases the lung anomaly was suspected prenatally. In 2 neonates the diagnosis of CCAM type III was only suspected postnatally.

Among the patients with CCAM type III intrauterine necrosis was stated 3 times, early deaths within 24 hours after delivery – 4 times, and late deaths – 2 times (1 on the 4th day of life, 1 on the 23rd day of life).

All patients with CCAM type III died before or after delivery.

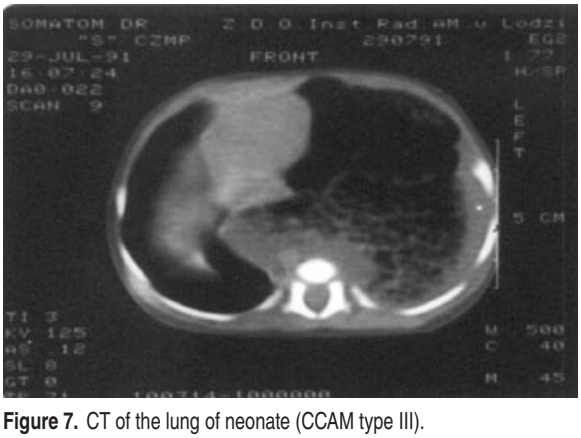

Figure 7. CT of the lung of neonate (CCAM type III).

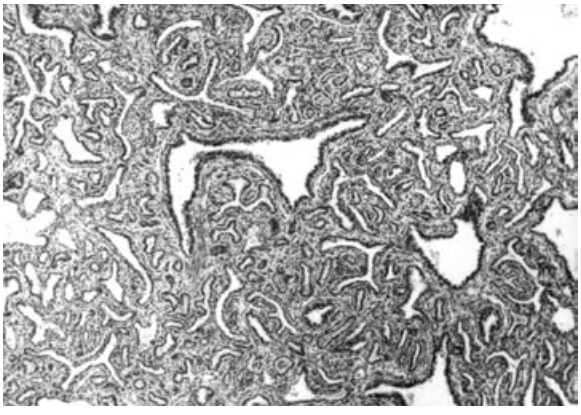

Figure 8. Pathology (CCAM type III) – bronchiolus – like spaces lined with flattened cuboidal epithelium.

Two patients with severe respiratory malfunctions were transferred to the intensive neonatal care unit of PMMHRI Department of Neonatology.

In one of the children, born in the 29th week of pregnancy, RDS was suspected in RTG of the abdomen, and pneumonia was described. CT was not performed. Pneumothorax requiring decompression occurred 3 times in the clinical course of the disease. In the second case – a full-term neonate born with birth weight of 3200 g and 3 points in the Apgar scale, a loud VSD-type murmur was heard above the heart. The pulmonary CT scan revealed developmental anomaly of the lungs with presence of cysts and displacement of the mediastinum (fig. 7). The clinical course was complicated with emphysema. Attempt of surgical treatment was made.

Both neonates died (first – on the 23rd day of life, second – on the 4th day of life), and the definite diagnosis was made on the basis of the result of anatomopathological examination (fig. 8).

Discussion

According to the analysis, congenital cystic adenoid malformation (CCAM) is a developmental anomaly which can be properly diagnosed and treated in the neonatal period [2, 7, 12, 13, 20].

The analysis of radiological criteria according to Stocker [10, 11] revealed that the most common radiological type of CCAM in children was type I.

In the radiological image diversity of sizes and wall thickness of the cysts were visible. The cysts contained air, rarely – only liquid, what made the image non-characteristic. Radiological exam in case of presence of multiple cysts might suggest the diagnosis.

The image of CCAM type I in the CT examination is typical. There are multiple septums in the lungs which present an image of thin-walled air ventricles (oval-shaped or roundish), well-circumscribed from the surrounding tissues, of different sizes (min 3 cm, max 10 cm). The mediastinal hernia is frequent, and the displacement of mediastinum was observed in 79% of cases.

The presented radiological material of our institute which comprises of a series of 8 neonates with CCAM type I, seems to corroborate these reports.

According to Stocker [10, 11], CCAM type II was a rarely considered radiological type as its radiological image is not characteristic.

Multiple small cysts, evenly located, contained liquid, air or both at the same time. The CT image of CCAM type II presented multiple oval, well-circumscribed, thin-walled areas of cystic type with the maximum diameter of 2.9 cm with the possibility of mediastinum and heart displacement.

In the presented group of 9 neonates with CCAM type II, the radiological images differed according to the size and localization of the cyst, and in most cases they were non-characteristic and required a definite confirmation based on the result of CT exam of the lungs. Other centers in the world have similar experiences [2, 5, 8, 11, 15, 21, 22].

The identification of cysts in the thorax in the course of CCAM in a neonate requires differential diagnostics, including diaphragmal hernia, bronchogen or enterogen cysts, intralobar and extralobar sequestration, congenital lobar emphysema, hypoplasia or agenesis of the lungs, pneumonia and RDS [1, 3, 4, 6, 7, 15, 21, 22, 23, 24].

According to Stocker, most neonates with the radiologically least common CCAM type III die after the delivery. Microcystic changes with the diameter of 0,5 cm, which constitute a homogenous mass, bilateral in most cases, are present in the lungs.

The CCAM type III was set on the basis of result of autopsy examination performed in our institute (Department of Clinical Pathomorphology – director: Professor A. Kulig)

Imaging diagnostics of the neonate, often with the use of CT, plays an important role in specifying the stated changes, especially in promising forms of CCAM. It relates to the assessment of type of the cyst, its size and location. The cysts can present as a homogenous mass or as separated cysts which cause the mediastinum displacement and lung compression [14, 20, 22, 24, 25]

Apart from selective CT examination of the lungs, echocardiac diagnosis and monitoring the state of a neonate can help in its further clinical assessment [7].

CCAM can be an isolated abnormality or coexist with other anomalies [3, 10, 22, 23, 26]. Among the significant anomalies accompanying CCAM, in the presented material intralobar sequestration was observed in one case (intraoperative histopathology examination), in the second case – bilateral pulmonary hipoplasia (autopsy exam), in the third – VSD-type interatrial septal defect (clinical and autopsy diagnosis).

According to the world literature [1, 2, 7, 10, 11] CCAM is limited to one lung in 80–95% of cases, but in 2% it can be a bilateral change. The analysis carried out by our institute corroborates this data. In 21 cases the anomalies were unilateral, while in the remaining 6 – bilateral. The changes affected both of the lungs in 5 cases of lethal form of CCAM type III, and in one case of promising CCAM type II.

Monitoring the CCAM gives the opportunity of respiratory support along with multidirectional treatment at INCU, provides the access to new techniques of radiological imaging diagnostics, early or planned surgical intervention on a neonate/child at the reference centre [6, 7].

According to our experiences, no matter what the clinical course of the disease was like (asymptomatic, hardly symptomatic, tachypnoe, acute respiratory failure), the neonate suspected with CCAM requires radiological imaging examinations (RTG of the thorax P-A, bok + CT of lungs).

The qualification for surgical treatment depends on the clinical course of the disease. Neonates with respiratory distress are operated at the earliest.

In order to exclude other anomalies it is suggested to carry out the transfontanelle USG, USG of the abdomen and ECHO of the heart.

Algorithm for proceeding in the neonatal period was proposed on the basis of our observations and Bagolan's data [2] and is depicted in fig. 9.

Conclusions

1. The suspicion of CCAM in the neonatal period requires monitoring at the reference centre.

2. The need for surgical treatment in CCAM neonates ought to be based on postnatal radiological diagnosis.

3. Full differentiating diagnostics is recommended in a neonate with CCAM.

4. Asymptomatic or hardly symptomatic neonate with CCAM can be difficult to identify on the basis of its clinical image and roentgen examination.

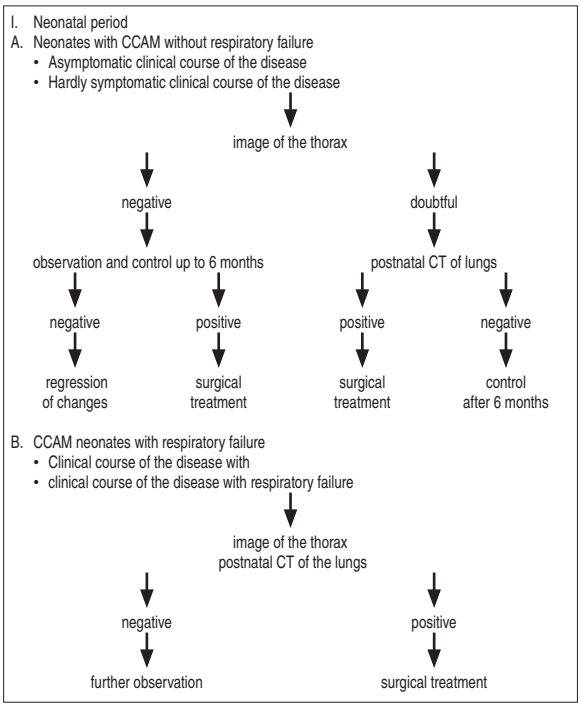

Figure 9. Algorithm of procedures for neonates with CCAM.

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