Ultrasonography in Children With Congenital Pyriform Sinus Fistula: Analysis of 31 Cases

Ultraschalluntersuchung bei Kindern mit angeborener Sinus-pyriformis-Fistel: Analyse von 31 Fällen

OPEN ACCESS

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ABSTRACT

Objectives Congenital pyriform sinus fistula (CPSF) is a rare disease that can be easily misdiagnosed. This study investigates the value of ultrasonography in the early diagnosis and treatment of CPSF in children.

Methods Clinical features and ultrasonography images of 31 CPSF pediatric patients confirmed by operation were retrospectively analyzed, different sonographic features during the infection period and the quiescence period were summarized and the consistency test of ultrasonic recognition and diagnosis between observers was conducted.

Results In this study, 25 CPSF children had thick-walled cystic masses during the infection period, and cystic masses of 8 cases showed gas echo inside; after the modified valsalva maneuver, gas echo was found in another 5 cases. The detection rate of gas can be enhanced through the modified valsalva maneuver and infants' cry so as to provide an important basis for the diagnosis of pyriform sinus fistula. During the quiescent period of inflammation of 6 cases, fistula can be completely shown, and the wall structure has not been completely destroyed, so that the running position of fistula can be clearly seen. Ultrasonography boasted a good inter-observer consistency in identification and determination (Kappa:0.799–0.857; $P < 0.001$).

Conclusion Ultrasonography could clearly reveal the position and direction of CPSF fistula. Different ultrasonic characteristics in different periods could provide relevant information for the selection of clinical operation timing and evaluate the post-operative effects.

ZUSAMMENFASSUNG

Zielsetzung Die kongenitale pyriforme Sinusfistel (CPSF) ist eine seltene Erkrankung, die leicht fehldiagnostiziert werden kann. In dieser Studie wird der Wert der Ultraschalluntersuchung bei der Frühdiagnose und Behandlung von CPSF bei Kindern untersucht.

Methoden Klinische Merkmale und Ultraschallbilder von 31 pädiatrischen CPSF-Patienten, die durch eine Operation bestätigt wurden, wurden retrospektiv analysiert, verschiedene sonografische Merkmale während der Infektions- und der Ruhephase wurden zusammengefasst und der Konsistenztest der Ultraschallerkennung und -diagnose zwischen Beobachtern wurde durchgeführt.

Ergebnisse In dieser Studie wiesen 25 CPSF-Kinder während der Infektionsperiode dickwandige zystische Massen auf, und die zystischen Massen von 8 Fällen zeigten ein Gasecho im Inneren; nach dem modifizierten Valsalva-Manöver wurde in weiteren 5 Fällen ein Gasecho gefunden. Die Gasnachweisrate kann durch das modifizierte Valsalva-Manöver und den Schrei des Säuglings erhöht werden, so dass sie eine wichtige Grundlage für die Diagnose der pyriformen Sinusfistel darstellt. In der Ruhephase der Entzündung kann in 6 Fällen die Fistel vollständig dargestellt werden, und die Wandstruktur ist nicht vollständig zerstört, so dass die Lauflage der Fistel deutlich zu erkennen ist. Die Ultraschalluntersuchung wies eine gute Übereinstimmung zwischen den Beobachtern bei der Identifizierung und Bestimmung auf (Kappa:0,799–0,857; P < 0,001).

Schlussfolgerung Mit Hilfe der Ultraschalluntersuchung konnte die Lage und Richtung der CPSF-Fistel eindeutig festgestellt werden. Unterschiedliche Ultraschallcharakteristika in verschiedenen Zeiträumen könnten relevante Informationen für die Auswahl des klinischen Operationszeitpunkts und die Bewertung der postoperativen Auswirkungen liefern.

Introduction

Congenital pyriform sinus fistula (CPSF) is a rare congenital branchial disease, whose fetal origin is controversial [1, 2]. The third branchial cleft deformity of CPSF originates in the basilar part of pyriform fossa, passes through the thyrohyoid membrane, winds upwards around the hypoglossal nerve and runs downwards until the lateral neck area in the downstream to end at the 1/3 of the posterior part of the front edge of sternocleidomastoid muscle, with the same course on both sides. The fourth branchial cleft deformity of CPSF originates in the apex of pyriform fossa, and runs downwards to the tracheoesophageal behind the thyroid gland to enter the chest; it is round the aortic arch in the left and round the subclavian artery to form the first loop;then, it goes upwards behind the arteria carotis communis to wind around the hypoglossal nerve to form the second loop; afterwards, it runs downwards to the lateral neck area to end at the 1/3 of the posterior part of sternocleidomastoid [3, 4]. It accounts for 1 %–4 % of all branchial malformations [5]. It is reportedly that 80 % of the cases occur in the childhood [6]. Surgery is the most effective therapy [7]. However, due to its rarity, it is often misdiagnosed and mistreated in clinical practise and the abscess incision drainage is performed, thus causing hyperplasia of scar tissues in the neck and normal anatomical structure destruction [8, 9]. This study aims to summarize the ultrasonographic manifestations of 31 CPSF cases and discuss the use of ultrasonographic features in early diagnosis and guiding treatment.

Materials and Methods

Case study

31 CPSF pediatric patients with complete data who were clinically diagnosed and underwent preoperative neck ultrasonography from October 2018 to October 2022 were selected. Inclusion criteria: ① Children under 18; ② patients and/or guardians who provide a written informed consent; ③ suspension laryngoscope reveals pyriform fossa to confirm the presence of internal orificium fistulae; ④ confirmed by open surgery. Exclusion criteria: Incomplete medical history provided by parents. Each pediatric patient's medical records, including demography, clinical manifestations and ultrasonic examination results are complete. This study has been approved by the Ethics Committee of our hospital.

Ultrasonography

Austria GE Volusion E8 and US GE Logic E9 ultrasonic diagnostic equipment, frequency of linear array probe 6 MHz-15 MHz, 18 MHz were adopted. With patients examined at supine position in a quiet or sleeping state, the equipment carefully scanned from the glottic level to hypothyroid level, observed the mass or fistula condition, carefully identified the running and opening position, measured and recorded the pipe diameter, length, and crossing position of fistula, and the mass size, boundary, internal echo and extension to the posterior cervical space. A retrospective diagnosis was made by two experienced sonographers of the images and finally analysis was made after their diagnosis results reached an agreement.

Statistical analysis

SPSS 20.0 software was used to carry out statistical analysis of the data. Qualitative data were expressed in numbers and percentages. Non-normally distributed quantitative data were expressed in medians, min and max values. Interobserver consistency was tested with kapp. Kappa value between 0.75 and 1 indicated good consistency; kappa value between 0.4 and 0.75 indicated medium consistency; kappa value between 0 and 0.4 indicated poor consistency; kappa value below 0 indicated extremely poor consistency and little practical significance.

Results

Clinical features of CPSF

In light of the research design, 31 CPSF pediatric patients confirmed by surgery and pathology enrolled in this study, including 19 male patients and 12 female patients, age 8–10 years old, median age 3 years and 6 months, duration of lesion 15 days to 3 years. Lesions were solitary in all cases, with 30 on the left side and 1 on the right side. Clinical features: thyroid cyst 25 cases, abscess of neck 4 cases, acute suppurative thyroiditis 5 cases, fever 15 cases, and restricted

▶ **Table 1** Summary of the Clinical Features of 31 Pediatric Patients With CPSF.

Clinical features	n	Proportion
Male (%)	19	61.29%
Female (%)	12	38.71%
Median age at first episode (range)	from 8 days to 10 years old	–
Median duration of the disease (range)	from 15 days to 3 years old	–
Left/right/bilateral	30/1/0	96.77%/3.23%/0
Clinical manifestation		
neck mass	25	80.65%
recurrent neck inflammation	4	12.90%
suppurative thyroiditis	5	16.13%
Fever	15	48.39%
restricted neck movement	2	6.45%
incision and drainage or puncture and pus aspiration	5	16.13%

neck movement 2 cases; incision and drainage of abscess was performed on 5 cases, as shown in ▶ **Table 1**.

Sonographic features of CPSF

Inflammatory infection period: 25 cases had cystic mass, irregular in 20, and regular in 5, wall thickness 3–5 mm; 15 had separated echo, and 2 had masses compressing adjacent airways, carotid arteries, and sternocleidomastoid muscles. Masses of 1 case extended to the retropharyngeal space. Cystic masses in 8 cases contained gas; after the modified valsalva maneuver, masses in 15 cases contained gas. With probe extrusion, 15 cases displayed liquid flow, as shown in ▶ **Table 2**. Color Doppler flow imaging (CDFI) revealed rich strip blood flow signals around the lesions (▶ **Fig. 1**)

Inflammation silent period: 6 cases displayed fistula, length 13–26 mm, median 20 mm, varying lumen thickness, bore size 1–5 mm; 2 cases had gas echo inside the lumen; fistulae of 3 cases travelled above the thyroid gland and those of another 3 travelled in the left intralobular thyroid gland. Color Doppler flow imaging (CDFI) revealed rich strip blood flow signals around the lesions (▶ **Fig. 2**)

There was excellent interobserver reliability for all the ultrasonic findings, ranging from 0.799 to 0.857, as shown in ▶ **Table 3**.

▶ **Table 2** Summary of the ultrasonic manifestation of 31 pediatric patients with CPSF.

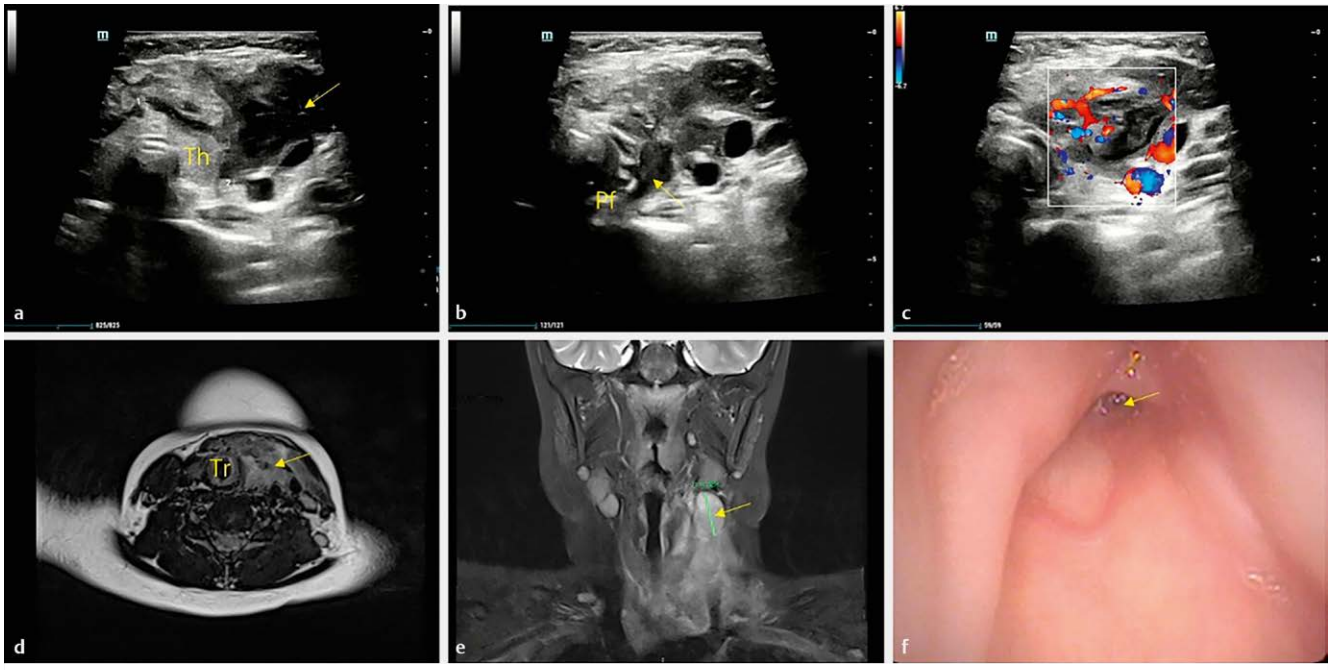
US finding		Value
Inflammatory infection period		
Neck position	Left/right/bilateral	14/1/0
Size (cm)		1.1–6.1 cm
Cyst wall		3–5 mm
Border	Well/poorly defined	4/11
Air bubbles in cysts	Yes/No	8/17
After the modified valsalva maneuver	Yes/No	15/10
Airway compression displacement	Yes/No	2/23
Into retropharyngeal space	Yes/No	1/24
Inflammation silent period		
Neck position	Left/right/bilateral	6/0/0
Fistula length		13~26 mm
Fistula diameter		1–5 mm
Running position	Beside the thyroid gland/ Inside the thyroid gland	3/3
Air bubbles in cysts	Yes/No	2/4
After the modified valsalva maneuver	Yes/No	2/4

Discussion

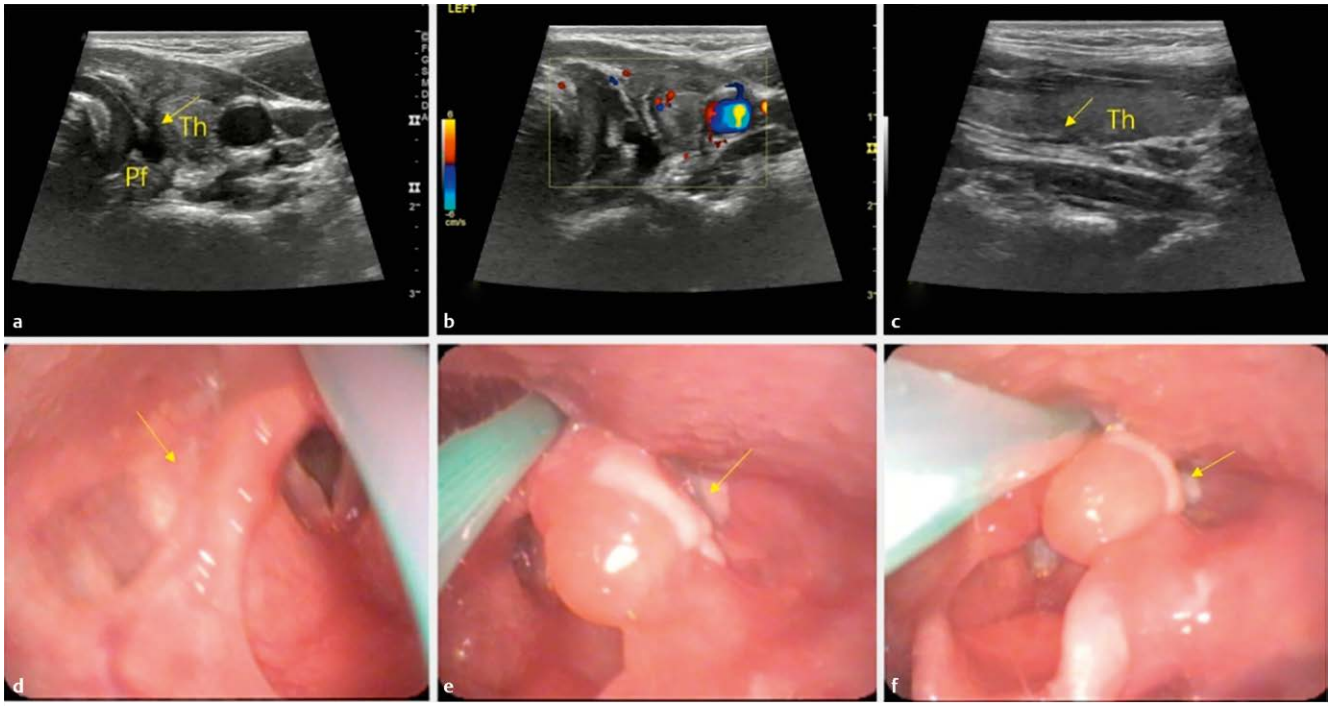
CPSF is a congenital branchial disease characterized with dormant onset, which can be detected only when the respiratory tract infection or oral infection occurs and bacteria enter the deep neck through the orificium fistulae to cause inflammation [10]. CPSF is widely regarded as fistula. Fundamentally, it is like a sinus tract with an internal os without an external os located in the pyriform fossa which only forms when the ruptured abscess penetrates the skin layer or in case of iatrogenic incision. Nearly all CPSF lesions are solitary and located in the left side [11]. In this group of cases, lesions are on the left side (96.77%), and on the right side (3.23%), with none on both sides. This may be related to the asymmetric development of the fourth branchial arch, the right branchial arch developing into the right subclavian artery and the left branchial arch developing into the arcus aortae. Some scholars believe that it is also related to factors such as the loss or degeneration of the right ultimobranchial body during embryonic stage [4]. Due to its special embryonic development and complex local anatomy make the early diagnosis of this disease challenging [12].

CPSF might occur at any age, which is more common in childhood. As previously reported in the literature [13], the first onset age is usually under 10, without gender differences, which is consistent with these research results. In this study, 4 cases (12.90%) were misdiagnosis as simple neck abscess, with the course of disease as long as 3 years. During this period, surgical exploration and incision and drainage of abscess were performed multiple times. 5 cases (16.13%) were misdiagnosed as acute suppurative thyroiditis and treated with puncture aspiration and anti-infection; the rest only received conservative treatment of anti-infection. These are not curative treatments for CPSF. Due to the persistence of fistula, secondary infection is easy to occur. Neonates and infants are characterized with weak development of tracheal cartilage. In case of infection, a larger cystic mass could easily appear, which may cause wheezing and acute respiratory distress. The prognosis will be good if it is resected completely as early as possible [14, 15].

Free from radiation and easy to operate, ultrasonography is the first choice for inspecting neck diseases. In this study, 25 cases of CPSF presented thick-walled cystic masses during the infection period, with dense punctiform and flocculent echo inside. It can be easily confused with other infectious lesions in the neck to lead to



▶ **Fig. 1** Inflammatory infection period of pyriform fossa. Male, 10, made hospital visits with a mass on the left cervical area without obvious cause, accompanied with pain and fever for more than one month. (**a**). A non-homogeneous echo mass appeared on the left cervical area with poorly-defined border, and much liquid dark was noticed (arrow); (**b**). the mass was in an irregular tubular shape extending towards the pyriform fossa (arrow); (**c**). abundant blood flow signals were around the mass; (**d**). the anterior part of axial T1WI left carotid sheath was mixed with signal mass shadow, and retropharyngeal space was not clear; (**e**). the inferior part of coronal T2WI left inferior parotid gland was mixed with signal mass shadow, with poorly-defined border; (**f**). ltaryngoscopy suggested suspected fistula in the left pyriform fossa, with secretion on the surface.



▶ **Fig. 2** Inflammation silent period of pyriform fossa. Male, 3 years and 9 months old, made hospital visits because of recurrent fever for 1 month. (**a**). The left cervical area showed strip parallel low echoes extending to the pyriform fossa, with clear boundaries (arrow); (**b**). no obvious blood flow signals were observed inside the strip parallel low echoes; (**c**). strip parallel low echoes went into the thyroid gland (arrow); (**d**). no obvious abnormality was found in the right pyriform fossa by the postoperative laryngoscope (arrow); (**e, f**). postoperative changes were discovered in the left pyriform fossa by the postoperative laryngoscope (arrow).

▶ **Table 3** Evaluation of the ultrasonic manifestation by two sonographers.

	Kappa	*P*
Neck position	0.799	< 0.001
Size (cm)	0.804	< 0.001
Border	0.854	< 0.001
Cyst wall	0.811	< 0.001
Fistula length	0.810	< 0.001
Running position	0.857	< 0.001
Fistula diameter	0.809	< 0.001

misdiagnosis. According to the previous literature, gas in the cystic mass is the characteristic sign of CPSF [16]. In this study, 8 cystic masses showed gas echo inside in routine examinations; after performing the modified valsalva maneuver, gas echo was found in another 5 cases. Previously reported in the literature, the detection rate of gas can be enhanced through the modified valsalva maneuver and infants' cry so as to provide an important basis for the diagnosis of pyriform sinus fistula [17].

In this study, during the quiescent period of inflammation in 6 cases, fistula can be completely shown given that the disease is caught early, fistula is slightly infected and the wall structure has not been completely destroyed. The deep running position of fistula in the pyriform fossa is restricted by surrounding tissues such as airway and retropharyngeal space. The fistula starts from the pyriform fossa and goes downwards to cross the thyroid parenchyma or enter parathyroid space along the posterior part of thyroid gland. Covered with an intact envelope, thyroid gland has abundant blood supply and is laden with iodine ions to inhibit the reproduction and growth of bacteria, so primary bacterial infection is difficult to occur. However, the fistula running has destroyed the normal anatomical structure and physiological characteristics of thyroid gland, and as a result, bacteria persistently invade adjacent gland tissues through the tube wall to give rise to secondary infection and to form thyroid abscess [18, 19]. Therefore, if the clinical feature is recurrent suppurative thyroiditis, it probably suggests the presence of potential dysplasia and CPSF should be highly suspected. High-frequency ultrasound can evaluate the infection degree of pyriform sinus fistula, which can provide important information for the selection of operation opportunity. In this study, two sonographers reached a good consistency in the identification and determination of ultrasound images, ranging from 0.799 to 0.857.

Limitations

This study has several limitations. First, the diagnostic accuracy of ultrasonography should be further evaluated, because the retrospective study is designed to merely include patients who are diagnosed with CPSF. Next, this study is to review the dynamic imaging, but the dynamic image evaluation has been restricted by the scanning range of the affected area of pyriform sinus fistula. Third, there are few neonates in this study, so the characteristics of pyriform sinus fistula in neonates cannot be summarized.

Conclusion

Sinusoid lesion between pyriform fossa and thyroid gland is often unnoticed or overshadowed by inflammation due to the deep position of sinus tract. For example, the gas in the sinus tract can better display the running direction of sinus tract; modified valsalva maneuver or infants' cry can help display the presence of gas. Ultrasonography can comprehensively evaluate the range of inflammation of the neck, thyroid gland involvement and serious complications. High-frequency ultrasonography not only can clearly display the presence and direction of fistula, but also can provide relevant information for the selection of clinical operation timing in light of different ultrasonic characteristics of CPSF in infectious and non infectious periods and evaluate the post-operative effects.

Contributor's Statement

L.Lei made substantial contributions to the conception and design of this study. W. Ruijie and L. Tingting and L. Zhouqin collected and reviewed the data. Z. Jingran and X. Haiyang made statistical analysis and computational analysis. L. Lei, Z. Qiuying and X. Fusui wrote the manuscript. W. Zebin and Z. Cailei and L. Zhou critically reviewed the manuscript and supervised the whole study process.

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Conflict of Interest

The authors declare that they have no conflict of interest.

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