PEDIATRIC INFERIOR VENA CAVA LESIONS: CLINICAL AND RADIOLOGICAL EVALUATION

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ABSTRACT

Objective: To investigate the value of multislice CT in detection and characterization of inferior vena cava lesions.

Subject and methods: This was prospective follow up study were done between Jaunary 2013 and May 2014 at Zagazig University Hospital. Patients selection was done in a simple random method. Patient inclusion criteria; the age ranging between 3-12 years of any sex, patients with IVC abnormality like abdominal enlargement and symptoms of liver disease or no symtoms with IVC lesions on CT abdomen. The study included 24 patients with a mean age of 7 ± 16.56 years. Patients were subjected to the following**:** Complete history taking**,** full clinical examination, full laboratory investigations, ultrasound with Doppler examination of the abdomen in all cases, multi-slice CT examination of the abdomen in all cases, conventional angiography when available.

Results: Final diagnosis was sex cases with Bud Chiari syndrome, four cases with retroperitoneal sarcoma, two cases with IVC leoimyosarcoma, three cases with retrocaval ureter, three cases with double IVC, two cases with left sided IVC and four cases with Wilms' tumor. MDCT shows accurate diagnosis in all patients in detection, characterization and differentiation of IVC lesions as proved with conventional venography and histopathological correlation. Ultrasound was important as initial screening method, but it was unable to characterize type of IVC thrombus in cases of Wilms' tumor.

Conclusion: Ultrasound is important as initial method for screening. Advantages of MDCT include superior spatial resolution; very fast image acquisition; and evaluation of adjacent and distant organs with one single imaging technique. MSCT angiography can be used as an alternative to US, MRI and DSA for evaluation of IVC lesions.

INTRODUCTION

The inferior vena cava can be a source for essential information for referring clinicians but often overlooked at abdominal imaging[(1)].

Congenital variations of the IVC are present in 0.3% to 0.5% of young, healthy adults. These occur as a result of abnormal embryologic development involving the vitelline, posterior cardinal, subcardinal, and supracardinal veins. They are present in approximately 4% of the population, and these individuals are often asymptomatic [2,3].

Because of anomalous persistence, regression, and anastomoses of the vitelline, posterior cardinal, subcardinal, and supracardinal veins, several classic congenital variations, including IVC absence or duplication, left-sided IVC, anomalous continuation of the IVC to the thorax, and retrocaval ureter, can occur alone or in combination [4,5].

Multislice computed tomography is considered to be the most common imaging that help to discover all inferior vena cava variants and IVC pathology because it is used to evaluate abdominal symptoms. CT angiography and CT with contrast with coronal and sagittal reconstructions detect the anomaly and help in deciding how to treat and determining the prognosis[6].

Advantages of multislice CT is good spatial resolution; short scan time and examination of nearby and distant organs[5]

The purpose of this work is to investigate the value of MSCT in detection and characterization of inferior vena cava lesions

PATIENTS AND METHODS

This is prospective follow up study were done between Jaunary 2013 and May 2014. This study included 24 patients with IVC lesions with or without signs and symptoms of IVC abnormality. Their ages ranging from 3-12 years with a mean of 7 ± 16.56 years, referred to the Pediatric and Radiology Department, Faculty of Medicine, Zagazig University Hospitals from the out patient clinics and Pediatric Surgery Department after obtaining institutional review board approval from our hospital and informed consent from patients before study.

Patient inclusion criteria; patients selection was done in a simple random method, their age ranging from 3-12years old and any sex, patients with IVC abnormality, and Patients with or without IVC lesion symptoms.

Patient exclusion criteria: patients with hypersensitivity to contrast agents, and patients with renal impairment.

Patients were subjected to the following: 1-Complete history taking.

2- Full clinical examination.

3- Full laboratory investigations.

4- Ultrasound with Doppler examination of the abdomen in all cases.

5- Multi-slice CT examination of the abdomen in all cases.

6- Conventional angiography when indicated, and

7- Operative findings and histopthological examination when available.

RESULTS

Table (1): Demographic data of the studied group:

Age (years)	**(n=24)**	
Mean ± SD	7 ± 16.56	
Range	3-12	
Sex	**No**	**%**
Male	18	75
Female	6	25

SD: *stander deviation*

This study included 24 patients. Their ages are ranging between 3 to 12 years, included 18 male (75%) and 6 females (25%).

Table (2): History and clinical data of the studied group:

Clinical data	**(*n*=24)**	
	No	**%**
Abdominal pain.	16	66.7
Abdominal enlargement	12	50
Symptoms of liver disease.	4	16.6
Asymptomatic (Accidental finding)	4	16.6

At this study abdominal pain (66.7%), and abdominal enlargement (50%) were the most common symptoms.

Table (3): Causes of Budd chiari syndrome

Cause	Number of patients	
Primary.	5	83.3%
Secondary to outside compression of hepatic veins.	1	16.6%

Cases of Budd Chiari was due to 1ry cause in 5 patients (83.3%) and 2ry to outside compression in one patient (16.6%).

Table (4): The CT findings in 6 cases of Budd-chiari syndrome:

CT findings	No. of cases	Percentage
Non opacification of hepatic veins and intrahepatic IVC	4	66.6
Enlarged caudate lobe	4	66.6
Decreased enhancement of peripheral portions of the liver and strong enhancement of central portions	4	66.6
Ascites	3	50
Calcified IVC thrombus	2	33.3
Intrahepatic comma-shaped collaterals	2	33.3
Multiple regenerative nodules	2	33.3

At this study non opacification of hepatic veins and intrahepatic IVC, enlarged caudate lobe and decreased enhancement of peripheral portions of the liver and strong enhancement of central portions were the most common CT findings in cases of Budd-Chiari syndrome.

Table (5): Aetiology of the studied group:

Final diagnosis	**(*n*=24)**	
	No	**%**
Budd Chiari syndrome	6	25
Large retroperitoneal sarcoma	4	16.6
Wilms' tumor	4	16.6
Retrocaval ureter	3	12.5
Double IVC	3	12.5
Left sided IVC	2	8.3
IVC leiomyosarcoma	2	8.3

The final diagnosis was made on basis of imaging and operative findings and histopathology. The most common cause of IVC lesions was Budd Chiari syndrome (6 cases) followed by retroperitoneal sarcoma and Wilms' tumor (each 4 cases).

Endoluminal recanalization and stent replacement in 5 patients with primary Budd- Chiari syndrome.

Femoral and jugular access venography, pressure measurement, ballon dilatation, metallic stent, control venography, and final pressure measurement were done.

Membraneous obstruction was seen in 3 patients and segmental obstruction in 2 patients.

Hepatic veins and IVC obstruction were seen in 4 patients and infrahepatic IVC obstruction in one patient.

Length of obstruction was ranging between 15-25mm, pressure measurement ranging between 10.2-20.6mmHg pre stenting and dropped between 4-14mmHg post stenting.

Retroperitoneal sarcoma

Four cases with retroperitoneal sarcoma were diagnosed with CT as large masses retroperitoneum with obvious compression of IVC.

Leiomyosarcoma

Two cases with leiomyosarcoma were diagnosed involving intrahepatic IVC, appearing as large soft tissue mass with cystic degeneration and calcification.

Wilms' tumor

Four cases with right sided Wilms' tumor were diagnosed with IVC tumor thrombosis.

Double IVC

Three cases with double IVC with retroaortic right renal vein and hemiazygos continuation of the IVC also shows thrombosis of intrahepatic IVC. They appeared as filling defect within the contrast filled IVC.

Retrocaval ureter

Three cases with retrocaval ureter were diagnosed in the study. A part of right ureter becomes trapped posterior and medial to the IVC, there was significant ureteric compression resulting in hydronephrosis with recurrent urinary tract infection.

Ultrasonography was valuable in detection of cases of IVC thrombosis ,while MDCT was superior to US in characterization and differentiation of nature of thrombosis, also MDCT was accurate method for detection of IVC anomalies and development.

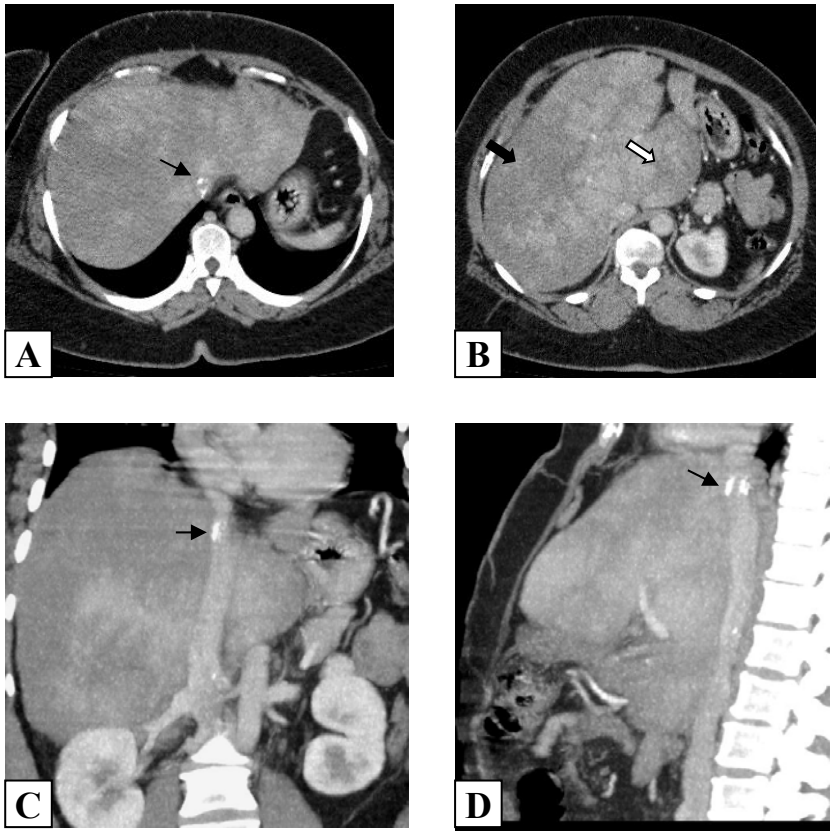
Histopathology was done in four cases including cases of wilm's tumor, retroperitoneal sarcoma, and two cases of IVC leiomyosarcoma.

Conventional venography was done in five cases of Budd chiari syndrome, three cases undergo ballon dilatation, two cases treated with stent placement.

Operative management was done in cases of wilm's tumor, and two cases of retroperitoneal sarcoma, other two cases were inoperable.

Case No (1): A case of 2ry Budd Chiari syndrome
Eleven years old female patient presented with abdominal pain at RT hypochondrium since 1year.
(A)& (B): Axial CT image;
- Intrahepatic IVC calcified thrombus (black arrow) (A).
- Enlarged caudate lobe with prominent enhancement (block white arrow) and decreased peripheral enhancement of right lobe (block black arrow) (B).

(C) & (D): Coronal and sagittal reformatted CT images;
- showing the calcified IVC thrombus (black arrow).



Case No. 2: A case of IVC leiomyosarcoma

Ten years female patient presented with vague abdominal pain and lower limbs edema since 6 month.

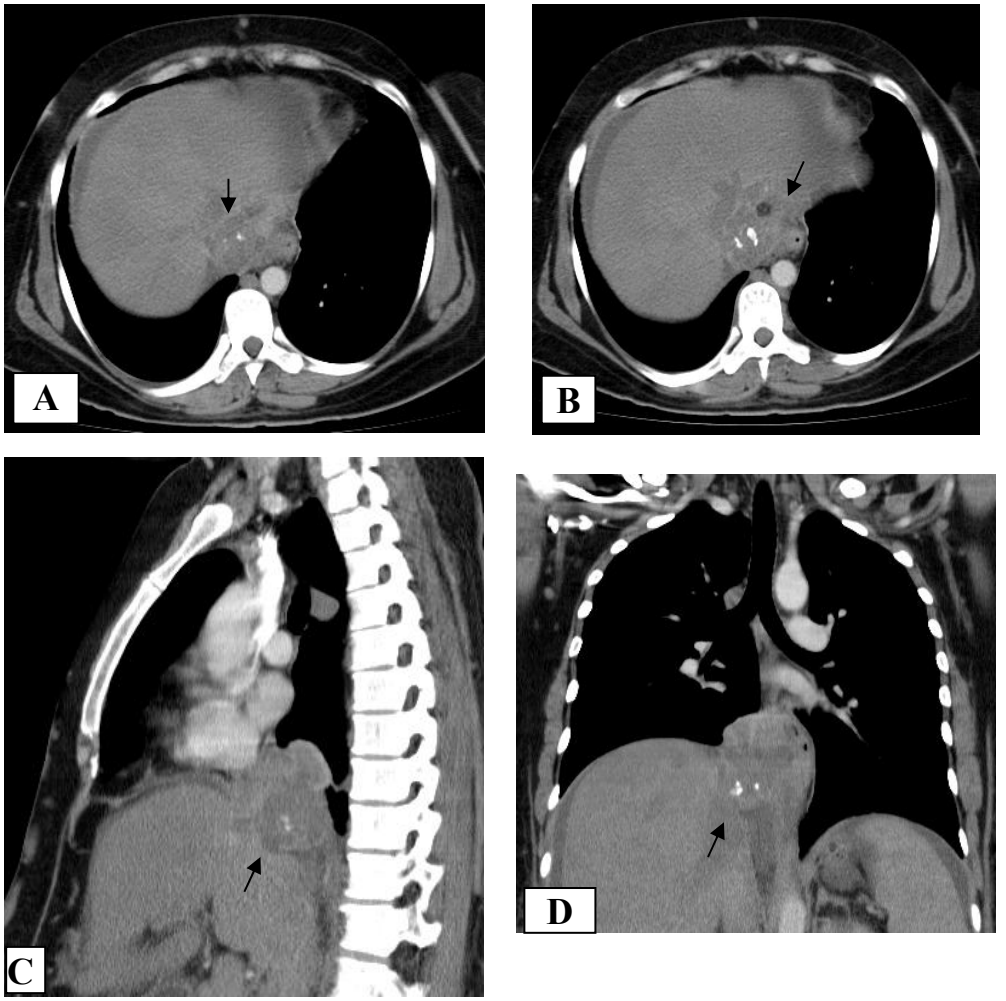
(A) & (B) : Axial MDCT images :

showing heterogeneous mass involving intrahepatic IVC with coarse calcification and cystic degeneration. Mass shows mild heterogeneous post contrast enhancemet. It measures 43 x 52mm and displacing esophagus with associated reticulation and haziness of intervening fat planes. (black arrows) associated with mild ascites.

(C) & (D) : Sagittal and Coronal MDCT reformatted images;

Illustrating good anatomical localization of IVC lesion with ascites (black arrow).

- Operative findings and histopathology confirmed the pathological diagnosis



Case No. 3: A case of 1ry Budd-Chiari Syndrome

Seven years old male presented with vague abdominal pain and abdominal enlargement.

(A) Abdominal Ultrasonography with Doppler;

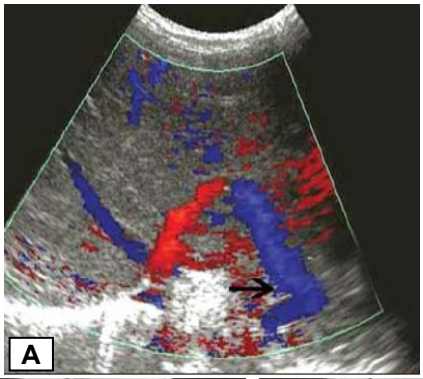
Capsular collaterals seen along the left subhepatic space (arrow) exiting through the liver in a transverse image through the left lobe. The middle hepatic vein is hepatofugal (blue), but the left hepatic vein is reversed (red) and flowing toward the large collateral (blue).

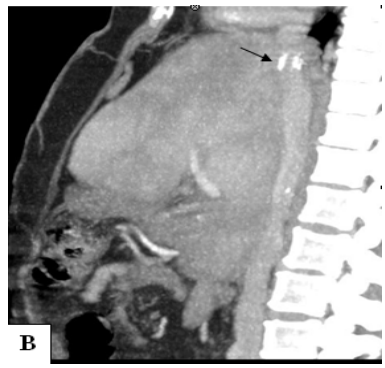
(B) A reformatted sagittal multislice CT image, reveals obvious narrowing of the IVC suprahepatic part.

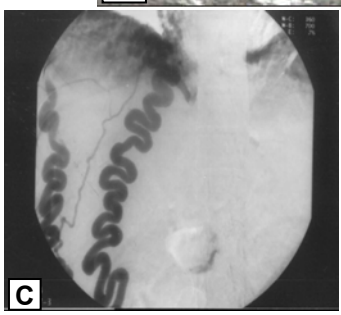
(C,D&E) transjugular and transfemoral venography, show lower and upper limit of IVC suprahepatic part obstruction with marked retroperitoneal collaterals bridging into superior surface of the liver.

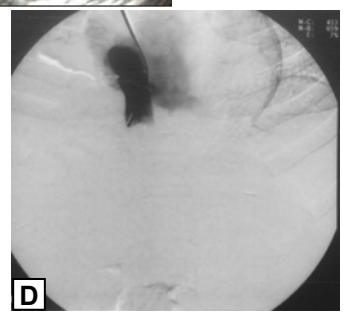
(F) the guide wire has passed the obstruction.

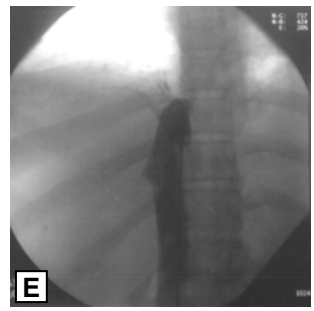
(G) balloon dilatation is shown and marked disappearance of the collaterals.

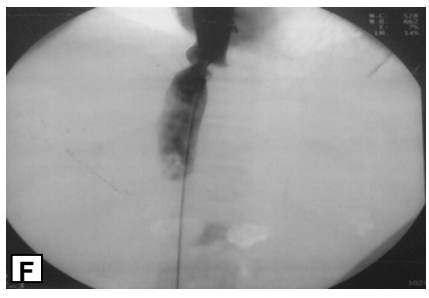


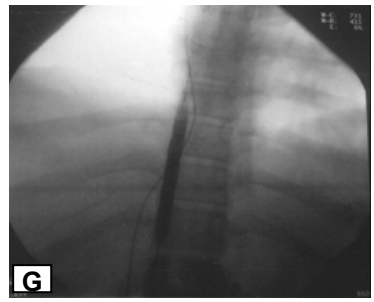












DISCUSSION

IVC is conduit of venous return to the heart from lower half of the body. The IVC is formed by the confluence of the right and left common iliac veins draining blood from the lower extremities and pelvis. As it ascends in the retroperitoneum to the right of the abdominal aorta, the IVC receives major tributaries including the lumbar veins, the left and right renal veins, the right gonadal vein, and the hepatic veins [(7)], so the lesions involving IVC can have great impact on the other systems of the body. We are trying to simplify the imaging approach on MDCT to classify these lesions into neoplastic and non neoplastic and also whether these arise primarily from IVC or as an extension of pathology involving other organs [(8).]

A basic knowledge of IVC embryogenesis is essential for understanding the anomalies of the IVC. The IVC is formed between the sixth and eighth gestational weeks by the sequential formation, anastomoses, and regression of three paired veins: the posterior cardinal vein, the subcardinal veins, and the supracardinal veins. It is composed of four segments: infrarenal, renal, suprarenal, and hepatic segments [(9)].

The right subcardinal vein forms the suprarenal segment of the IVC, and the right supracardinal vein forms the infrarenal segment. Anastomotic channels between the supracardinal and subcardinal veins form the intervening renal segment of the IVC. Cranially, the subcardinal vein (suprarenal IVC) forms an anastomosis with the hepatic segment, which is derived from the right vitelline vein. Caudally, the infrarenal IVC (right supracardinal vein) forms an anastomosis with the iliac veins[(10)].

Given the complex embryogenesis of the IVC, it is not surprising to encounter anatomic variations. Duplication of the IVC occurs in up to 2.8% of the population. Failure of the left caudal supracardinal vein to regress results in persistent communication between the left common iliac vein and the left renal vein [(11)].

Though asymptomatic, double IVC has important clinical implications when attempting caval filtration or when retroperitoneal surgery is performed. The left IVC can also be misdiagnosed as lymphadenopathy on cross-sectional imaging [(11)].

The left IVC typically ends at the left renal vein, which crosses

anterior to the aorta in the normal fashion to join the right IVC. However, there may be variations in this arrangement "Double IVC with Retroaortic Right Renal Vein and Hemiazygos Continuation of the IVC" [12].

Once a true filling defect of the IVC is established, identification of the cause, whether benign or malignant, and extent will guide clinical treatment [14].

IVC masses can be classified into benign and malignant pathologies. Leiomyomatosis come under benign masses [13].

Leiomyosarcoma is the most common primary malignancy involving the IVC .

Leiomyosarcoma arises from the smooth muscle cells in the vessel wall. The initial growth of an IVC leiomyosarcoma is intramural. Two-thirds of tumors will demonstrate predominantly extraluminal growth, and one-third will demonstrate predominantly intraluminal growth. The intraluminal tumors may cause venous obstruction [15].

Leiomyosarcomas of the IVC are classified according to their location. Approximately 37% of tumors occur in segment I, below the level of the renal veins and above the iliac vein bifurcation; 43% involve segment II, between the renal veins and the level of the hepatic veins; and 20% form in segment III at or above the hepatic veins level and may extend into the right atrium. The level of IVC involvement is important because tumors involving the renal and suprarenal IVC are associated with the most favorable prognosis[16].

Involvement of the intrahepatic IVC is associated with the worst prognosis.

At imaging, the predominantly intraluminal leiomyosarcoma focally dilates and usually obstructs the IVC and may display homogeneous contrast enhancement exophytic leiomyosarcomas appear as large retroperitoneal masses with heterogeneous contrast enhancement. Cystic necrotic areas are not rare.

In this work we diagnosed two case as IVC leiomyosarcoma, in which CT revealed an ill defined heterogeneous enhanced retroperitoneal mass with multiple areas of cystic necrosis and calcifications involving segment III of the IVC and that the adjacent abdominal organs were normal with no evidence of other abdominal tumors[15].

Budd-Chiari syndrome is applied to the clinical manifestations of hepatic venous outflow obstruction at any level from the

small hepatic veins to the junction of the inferior vena cava and the right atrium regardless of the cause of obstruction. Early diagnosis of Budd-Chiari syndrome is important for establishing appropriate treatment. Therefore imaging studies combined with clinical information are often essential for reaching a definitive diagnosis [17].

Contrast-enhanced CT scan findings in patients with Budd-Chiari syndrome include an inhomogeneous mottled liver with delayed enhancement in the periphery of the liver and around the hepatic veins[18].

The peripheral zones of the liver may appear hypoattenuating because of reversed portal venous blood flow, which results from increased postsinusoidal pressure produced by hepatic venous obstruction.

The caudate lobe is enlarged (caudate lobe hypertrophy) and demonstrates increased contrast enhancement compared with the remainder of the liver. Identification of hepatic veins may fail. Thrombosis within the hepatic veins and the IVC can be identified in 18-53% of patients[19].

Our criteria for diagnosing Budd-Chiari syndrome were bases on the study of **Brancatelli et al., 2007,** who stated that, the CT appearance of Budd-Chiari syndrome is well recognized, in acute Budd-Chiari syndrome, occlusion of the hepatic veins with ascites is the typical finding. The liver exhibits patchy, decreased peripheral enhancement caused by portal and sinusoidal stasis and stronger enhancement of the central portion of the liver parenchyma and the inferior vena cava is compressed by the enlarged caudate lobe. In subacute or chronic Budd-Chiari syndrome, portosystemic and intrahepatic comma shape collateral vessels are often found. Contrast-enhanced CT is useful for depicting regions of hypoperfused liver parenchyma. Low-attenuation thrombus may be visible in the hepatic veins and IVC. Chronic thrombosis of the inferior vena cava can evolve into calcification.

In this study, We diagnosed sex cases as Budd-Chiari syndrome, hypoattenuattion of the hepatic veins was found in four cases, ascites was found in 3 cases, decreased enhancement of peripheral portions of the liver and strong enhancement of central portions was found in 4 cases, so reported as acute Budd-Chiari syndrome. Enlarged caudate lobe

found in 4 cases. In two cases calcified IVC thrombus was detected, so reported as chronic Budd-Chiari syndrome, 2 cases had associated intrahepatic comma shape collateral vessels [20].

Chronic Budd-Chiari syndrome is also characterized by the development of multiple regenerative nodules. These nodules are usually multiple and have a diameter of 0.5–4.0 cm. On multiphasic CT, regenerative nodules are markedly and homogeneously hyperattenuating on arterial phase images and remain slightly hyperattenuating on portal venous phase images [20].

That was in agreement with our study as we found multiple regenerative nodules in two cases of chronic Budd-Chiari syndrome, with diameter of 10-18 mm, homogenously enhanced in arterial and portovenous phases).

We concluded that MDCT can adequately detect pathologic conditions affecting the IVC. Advantages of MDCT include superior spatial resolution; very fast image acquisition; and evaluation of adjacent and distant organs with one single imaging technique, which is critical in patients with tumor involving the IVC[20].

In conclusion, ultrasound is important as initial method for screening. Advantages of MDCT include superior spatial resolution; very fast image acquisition; and evaluation of adjacent and distant organs with one single imaging technique.

Multi–detector row CT angiography can be used as an alternative to US , MRI and DSA for evaluation of IVC lesions.

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دور الفحص الاكلينيكي والتصوير المقطعي المتعدد الشرائح في تقييم افات الوريد الاجوف السفلي بالأطفال

الهدف من البحث: تقييم دور الفحص بالأشعة المقطعيه متعددة الشرائح في اكتشاف وتقييم افات الوريد الاجوف السفلي بالأطفال.

المرضي وطرق الفحص: شملت هذه الدراسة 24 مريضا (18 ذكر و6 إناث) تتراوح اعمارهم بين 3-12 عاما وتم الفحص الاكلينيكي والمعملي والموجات الفوق الصوتية بالدوبلر الملون والأشعة المقطعية متعددة الشرائح لجميع المرضي.

نتائج البحث: كان التشخيص النهائي 4 حالات تخثر في الوريد الاجوف السفلي نتيجة ورم ويلمز بالكلي ،ست حالات متلازمة بد كياري، وأربع حالات ساركوما خلف الصفاق، حالتين تعاني من ساركوما الوريد الاجوف السفلي، ثلاث حالات للحالب خلف الأجوف وثلاث حالات لازدواج الوريد الاجوف السفلي، وحالتين الوريد الاجوف علي الجهة اليسري. اظهرت الأشعة المقطعيه متعددة الشرائح التشخيص الدقيق في جميع المرضى في كشف وتوصيف وتصنيف الآفات.

خلاصه البحث: الموجات فوق الصوتيه تعتبر طريقة الفحص الأولية.

تعتبر الأشعة المقطعيه متعددة الشرائح بالصبغة علي الوريد الاجوف السفلي ذات قيمه في تشخيص وتصنيف و تحديد مدي انتشار افات الوريد الاجوف السفلي وبالتالي توجيه طريقه العلاج و يمكن الاستعاضة بها عن الفحص الوريدي بالقسطرة .