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RESEARCH ARTICLE

CONGENITAL CAROTID-JUGULAR FISTULA IN A 7-MONTH-OLD INFANT: RARE CASE AND LITERATURE REVIEW

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Abstract

Carotid-jugular fistulas (CJF) are extremely rare and serious vascular anomalies, especially in infants. Despite their rarity, these fistulas can cause significant complications. We present the case of a 7-month-old infant diagnosed with a congenital CJF, emphasizing the diagnostic challenges and treatment options for this uncommon condition in young patients.

Introduction:-

Congenital carotid-jugular fistulas, rare anomalies, represent congenital vascular malformations characterized by a connection between the external carotid artery and the internal jugular vein, resulting from multiple disturbances occurring at various stages of embryonic development. This anomaly progresses gradually and can remain asymptomatic until it triggers symptoms such as localized swelling and manifestations of an arteriovenous shunt [1]. Although several cases of this malformation have been reported in the literature [2], there is no clear consensus regarding the optimal surgical approach. We present here a recent case of a congenital carotid-jugular fistula in a 7-month-old patient, successfully treated surgically without complications. To our knowledge, this is the first documented case of CJF in this age group reported in the medical literature. It is imperative to seek a balance between maximum therapeutic efficacy and minimal risk, promoting early management to prevent potential severe complications

Patient and Observation:-

Infant SS was admitted to the intensive care unit of the Oujda University Hospital Center on 11/20/2022 due to intractable vomiting following severe gastroenteritis complicated by acute dehydration requiring vascular access. An ultrasound-guided attempt to place a central venous line was made, but suspicion of an arteriovenous anomaly led to the placement of the central line on the contralateral (left) side. After stabilizing the child, additional examinations including a Doppler ultrasound and a comprehensive cardiac evaluation were performed, leading to the diagnosis of a carotid-jugular arteriovenous fistula (CJF), establishing a communication between the left internal jugular vein and the left common carotid artery. An opening of 2.7 mm was discovered on the wall of both vessels (see Figures 1 and 2), with a recording of arterialized flow in the vein, without neurological deficits or signs of heart failure.

The evaluation report of infant S.S. highlights a normal cardiovascular status, without signs of respiratory distress. Cardiac auscultation reveals no audible murmurs, and the electrocardiogram shows normal cardiac activity with a heart rate of 148 beats per minute.

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Cardiac ultrasound measurements reveal overall normal dimensions of the left ventricle, posterior wall, and septum. No right ventricular dilation is observed, and septal kinetics are deemed normal. The mitral and aortic valves also exhibit normal kinetics. Finally, the parameters of systolic function indicate overall satisfactory cardiac performance, with a shortening fraction of 40% and an ejection fraction of 78%. Despite the presence of the cervical arteriovenous fistula, the cardiovascular status of infant SS remains overall stable and does not show signs of major cardiac dysfunction (see Figure 3).

An endovascular surgical intervention was then performed to treat the CJF, involving the excision of the fistula after proximal and distal control of the external carotid artery and internal jugular vein on either side of the communication. Postoperative follow-up revealed no neurological or cardiac complications. The patient was reviewed at three and six months with a follow-up cervical Doppler ultrasound, which showed no abnormalities

Discussion:-

Arteriovenous fistulas (AVF) in the head and neck region are infrequent. Most AVFs are of traumatic origin or secondary to iatrogenic causes [1]. Congenital carotid-jugular fistulas (CJF) are rarer but more serious vascular anomalies (4%) [2][3][4]. Reports concerning congenital AVFs involving the common carotid system are much fewer, with only a little over 20 cases documented to date [5]. These fistulas are characterized by an abnormal acquired communication between the carotid artery and the internal jugular vein. Although uncommon, these fistulas can lead to significant complications in infants [4]. Research indicates that congenital arteriovenous fistulas located in the head and neck are typically unilateral[6]. A congenital arteriovenous fistula is a morphologically benign lesion, but it tends to grow rapidly in clinical practice and must be monitored [6][7][8]

Most of the time, arteriovenous communications involving the major vessels of the neck are caused by trauma (37%), predominantly affecting males (67%) [10], whether they are blunt or penetrating, or may occur following surgical intervention or extensive infection of the neck region [11]

Unlike arteriovenous fistulas in other parts of the body, those affecting the carotid-jugular region are particularly prone to complications such as refractory high-output heart failure and atrial fibrillation. Given that the carotid artery is a direct branch of the aorta, when a carotid arteriovenous fistula occurs, a large amount of carotid blood flows rapidly into the jugular vein through the fistula and leads to increased venous pressure [12],increased blood flow to the heart, and rapid clinical decompensation, which can result in cardiac enlargement. Progressive cardiac enlargement can lead to heart failure, and carotid arteriovenous fistulas may appear early and cause severe heart failure [13]. Even though they are rare, carotid-jugular fistulas should be considered in the differential diagnosis of cervical masses [14].

Typically, in patients presenting with a carotid-jugular fistula, the compression of the carotid artery serves to halt the characteristic bruit [15]. The Branham sign, indicative of bradycardia and elevated mean arterial pressure upon compression to impede blood flow across the fistula, may be discerned [16]. Herein, we encapsulate post-2017 published endeavors (Table 1). The clinical manifestations of congenital AVFs hinge upon the magnitude of the shunt. Foremost among these is a pulsatile mass, although ancillary symptoms such as a thrill, a murmur, pulsatile tinnitus, cephalalgia, vertigo, or discomfort may likewise manifest [17, 18, 19, 20, 21]. Prolonged neglect of the fistula can yield manifold complications encompassing heart failure, fistula rupture, or embolic sequelae [20, 21]. Furthermore, the protracted existence of the fistula can precipitate a steal phenomenon. The subject of our inquiry was a 7-month-old neonate with an asymptomatic fistula. Graced by fortuitous and prompt malady detection, the patient evinced no cardiac or neurological sequelae, exhibiting a normative cardiac ejection fraction.

Tableau 1 : Summary of Literature Post-2017 Regarding Congenital Carotid-Jugular Fistula

Author	Age (year)/Sex	Clinical presentation	Artery concerned	Diagnostic Imaging	The technique used	Résultat	Impact
Deqiu Cui et al,2017[17]	***9/M***	***Pulsatile mass and thrill***	***ECA and the EJV.***	DSA	Spiral embolization	**CO**	**NC**
Bellosta et al, 2017[18]	***15/F***	***Pulsatile mass, a murmur, dizziness***	**ECA and the IJV.**	CTA	**Spiral embolization**	**CO**	**NC**
Rong Xu Du et al, 2017[19]	***13/ M***	***Pulsatile mass, cardiac dilatation, pulmonary hypertension, and thrill***	The bilateral external carotid artery and IJV	CTA	Endovascular surgery	**CO**	***NC***
Ming Wang et al, 2018[20]	***64/F***	**Pulsatile mass and right hemiparesis following a cough**	**-**	DSA	**Fractionated embolization assisted by stent.**	**CO**	**NC**
Jia Hu et al, 2019[21]	***27 / semaines de grossesse***	**Massive tricuspid regurgitation in the fetal heart, complete cardiac hypertrophy.**	**-**	**Échodoppler**	The pregnant patient underwent a termination of pregnancy at 27 weeks.		
Notre cas	***7mois /M***	***Asymptomatic***	***Artere carotide externe et l artere jugulaire interene***	**Échodoppler**	Embolization	**CO**	NC

M *male,* ***F*** *female,* ***DSA*** *subtraction digital angiography,* ***CTA*** *computed tomography angiography, ,* ***ECA*** *external carotid artery,* ***IJV*** *internal jugular vein,* ***EJV*** *external jugular vein,* ***NC*** *No complications ,****CO*** *Complete occlusion*

The diagnosis of the disease is not difficult; according to data gathered in the literature, arteriography accounts for 45% of confirmatory examinations [22], followed by CT angiography at 25% and Doppler ultrasound at 23% [23]. The utilization of MR angiography is less common, at only 7% [24]. MR angiography is seldom performed, but it may be beneficial when endovascular treatment is considered [25].

Two-dimensional ultrasound can be challenging, but color Doppler ultrasound serves as a crucial diagnostic tool for arteriovenous fistulas. This technique reveals blood flow from the carotid artery passing through the fistula and into the jugular vein, showing turbulent flow with color artifacts at the site of the fistula, thus confirming the presence of high-velocity arteriovenous communication. In this case, color Doppler ultrasound displayed blood flow signals within the abnormal echogenic area (see Figure 1). The left carotid artery and left internal jugular vein exhibited color artifacts at the distal end and an arterialized waveform within the vein[26].

The objective of therapy aims at fistula closure while preserving the integrity of the main arterial pathway, achievable through either surgical intervention entailing fistula excision followed by vascular wall reconstruction, or an endovascular modality [27]. Consideration can be given to either surgical or endovascular management for CJF. The latter approach may involve the utilization of detachable balloons, a method perceived as safer and less invasive. The preference for employing detachable balloons and coils in treating congenital AVFs persists, notwithstanding their propensity for recurrence. The intricate and extensive ramifications of AVFs render their complete eradication challenging, with complete remission improbable.

Congenital cervical-jugular fetal AVFs can lead to congestive heart failure and fetal demise, underscoring the crucial importance of accurate diagnosis for maternal and fetal health [21]. The majority of reported cases have undergone surgical intervention with various approaches depending on the extent and location of the lesion [8,15], either through surgery (34%) or endovascular methods (58%) [17][20]. Unoperated patients were rare (8%) [3][9].

Open surgical repair, despite its reduced frequency following the emergence of endovascular techniques, maintains its significance as a safe and efficacious therapeutic modality for this rare condition, owing to the accessibility of cervical vasculature and lower incidence of thromboembolic complications [15][17], although the superiority between approaches remains debated [15][18]. Nonetheless, irrespective of the treatment modality employed, diligent patient monitoring is paramount to promptly identify any signs of aneurysmal degeneration [26].

It is imperative for physicians to carefully examine newborns and infants to promptly screen for this serious condition, enabling tailored management and significant improvement in clinical outcomes [26][28].

Figure:

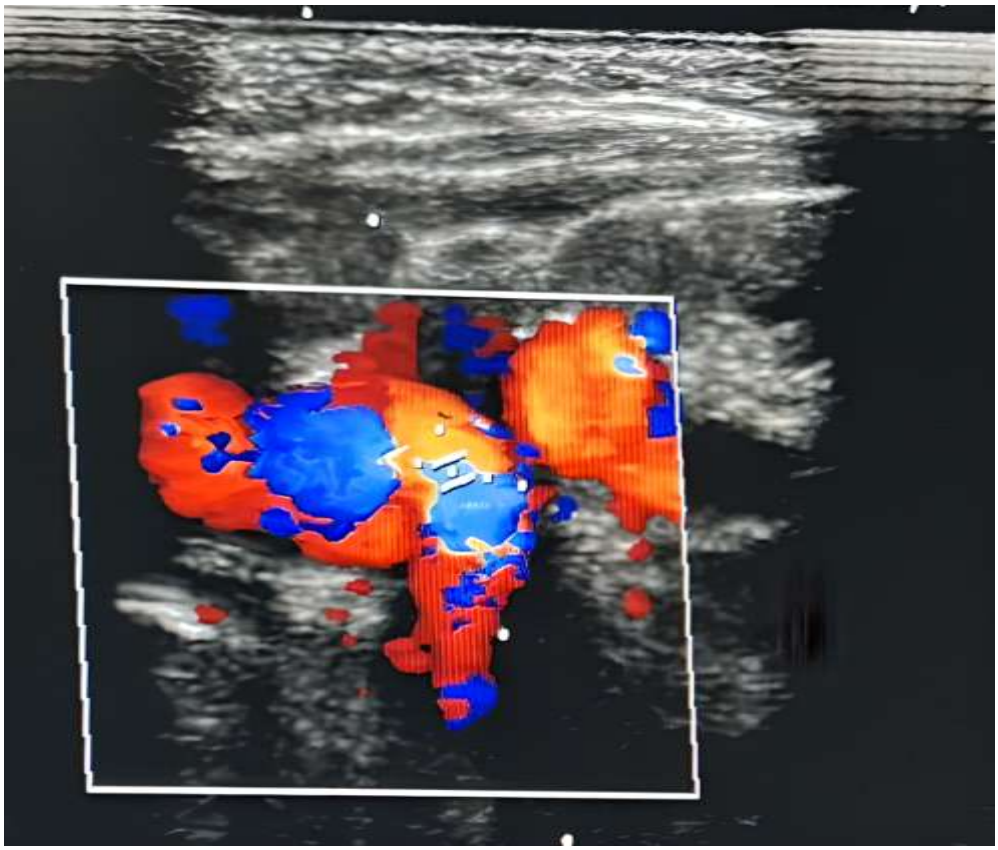

Figure 1:- Color Doppler echocardiogram image showing direct communication between the external carotid artery and the internal jugular vein.

It is observed that the left carotid artery and left internal jugular vein are connected, with the spectrum of arterial blood flow detectable within the venous vessel.

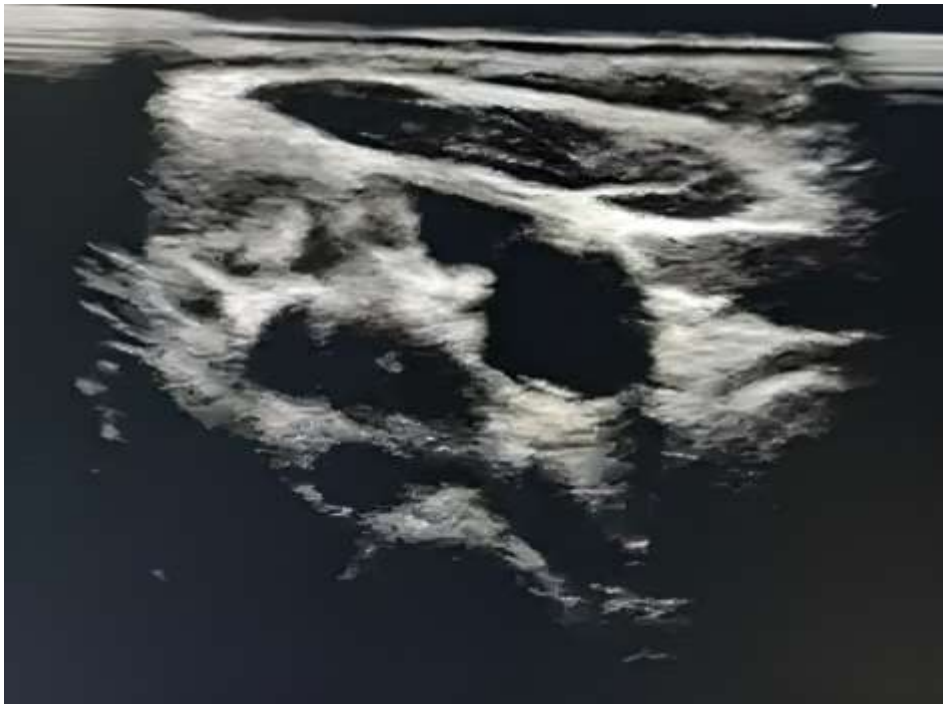

Figure 2:- Image of cervical Doppler ultrasound showing a large carotid-jugular fistula with a diameter of 2.7 mm between the external carotid artery and the internal jugular vein.

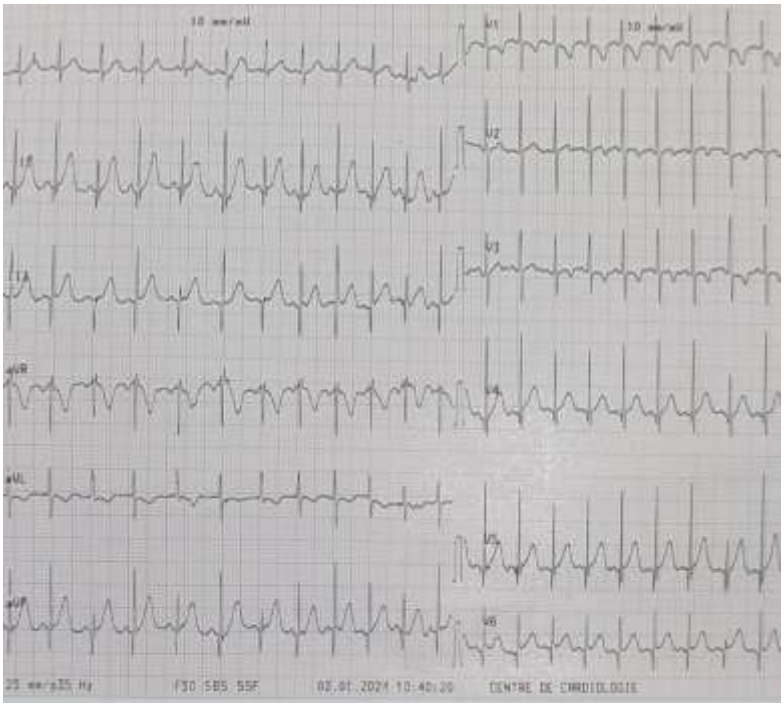

Figure 3:- Normal Electrocardiogram: Physiological respiratory arrhythmia with a heart rate of 148 beats per minute. - QRS axis at +60°. - PR interval at 8/100 seconds. - Normal right ventricular hypertrophy for age. - No recorded arrhythmia.

Conclusion:-

In conclusion, carotid-jugular fistula (CJF) is a rare complication that can be serious and may occur congenitally, iatrogenically, or traumatically. The promptness of diagnosis and treatment is crucial to avoid potentially fatal complications such as congestive heart failure or cerebral thromboembolic events

Conflict of Interest:
The authors declare no conflicts of interest.

Authors' Contributions:
All authors contributed to the conception and design of the study, data collection, and writing of the manuscript. All authors have read and approved the final version of the manuscript.

References:-

1. Kohout MP, Hansen M, Pribaz JJ, Mulliken JB (1998) Arteriovenous malformations of the head and neck: natural history and management. PlastReconstr Surg 102:643–654
2. Florine Randrianambininjanahary, S. M., Randriantianarisoa, K., Gabrielle Andrianah, E. P., Hasina Rajaonarison, N. O. N. L., & Ahmad, A. (2022). Fistule carotido-jugulaire post-traumatique: à propos d'un cas. Sang Thrombose Vaisseaux, 34(6)
3. Raherinantenaina, F., Avotsihoarana, T. H., Rabemanorintsoa, F., Rabemanantsoa, R., Andrianandraina, M. C. G., Ahmad, A., ... &Rajaonanahary, T. M. A. (2019). Prise en charge des fistules carotido-jugulaires: cas clinique et revue de la littérature. JMV-Journal de MédecineVasculaire, 44(6), 426-431.
4. J.Y. Ahn et al. Stent-graft placement in a traumatic internal carotid-internal jugular fistula and pseudoaneurysm J Clin Neurosci (2004)
5. Faghihi LT, Arjmand SA, Pourghorban R, Khalili PE (2015)Congenital external carotid-external jugular arteriovenous fistula: diagnosis with contrast-enhanced computed tomography. Iran J Radiol12:e7450 literature. Neurosurgery 30:272–276
6. Bellosta R, Vescovi M, Attisani L, et al. Endovascular treatment of congenital external carotid-jugular fistula: case report and review of the literature. Vasc Endovascular Surg 2017;51:316–9.
7. Li JC, Cai S, Jiang YX, et al. Diagnostic criteria for locating acquired arteriovenous fistulas with color Doppler sonography. J Clin Ultrasound 2002;30:336–42.
8. Zahdi, O., Lahlou, N. D., Bhali, H. E., Hormat-Allah, M., Khloufi, S. E., Sefiani, Y., ... &Lekehal, B. (2020). Iatrogenic carotid-jugular arterio-venous fistula: a rare complication associated with jugular venous catheterization (a case report). The Pan African Medical Journal, 37, 379-379.
9. Wadhwa R, Toms J, Nanda A, Abreo K, Cuellar H. Angioplasty and stenting of a jugular-carotid fistula resulting from the inadvertent placement of a hemodialysis catheter: case report and review of literature. Semin Dial. 2012 Jul;25(4):460-3 Epub 2011 Dec 9
10. Alejandro Santillan , Jeremiah Johnson and Lee A Birnbaum, Embolization of a congenital arteriovenous fistula of the internal maxillary artery: A case report and review of the literature, nterventional Neuroradiology 2016, Vol. 22(3) 349–353! The Author(s) 2016
11. Chan CC, Lee V, Chu W, Tam YH, Li CK, Shing MM. Carotid jugular arteriovenous fistula: an unusual complication of internal jugular vein catheterization in children. Pediatr Blood Cancer 2012; 59:1302—4
12. Dammak A, Ben Jmaà H, Hadhri S, Cheikhrouhou H, Dhouib F, Haddar S, et al. post-traumatic carotido-jugular fistula: case report and review of the literature. J Med Vasc 2017; 42:388—9
13. Nandapalan V, O'Sullivan DG, Siodlak M, Charters P. Acute air-way obstruction due to ruptured aneurysmal arterio-venous fistula: common carotid artery to internal jugular vein. J Laryn- golOtol1995;109:562—4
14. Caldarelli, C., Biricotti, M., Materazzi, G., Spinelli, C., &Spisni, R. (2013). Acquired carotid-jugular fistula: its changing history and management. International Scholarly Research Notices, 2013.
15. Mlynski A, Mocellin N, Imperato M, Molimard B, Bordier E, Menguy P, et al. Treatment of a ballistic wound of the com- mon carotid artery revealed by a spontaneous carotid-jugular fistula. Ann Vasc Surg 2011;25:269 [e9-12].
16. Huang CF. Pulsatile tinnitus from a spontaneous carotid-jugular fistula: case report. Neurologist 2009;15:335—7
17. Chen JY, Chan SH, Lin LJ, Luo CY. Late-onset congestive heart failure with multiple carotid-jugular fistulae and pseu- doaneurysm after penetration injury. J Formos Med Assoc 2006;105:844—7
18. Kim BS, Lee SK, terBrugge KG. Endovascular treatment of congenital arteriovenous fistulae of the internal maxillary artery. Neuroradiol2003;45:445—50.
19. Lozman H, Nussbaum M, Yang W. Planned carotid artery jugular vein fistula: 30-year follow-up and surgical correction. Arch Surg 1982;117:1343—5
20. DeqiuCui ,Jingwei Li ,Gao Zeng ,Xinglong Zhi &Jianxin Du , A congenital external carotid artery-external jugular vein arteriovenous fistula was successfully treated by coil embolization (case report and literature review) , 2017 DOI: 10.1007/s00381-017-3488-9

21. Raffaello Bellosta , Monica Vescovi , Luca Attisani , Luca Luzzani , Endovascular Treatment of Congenital External Carotid-Jugular Fistula: Case Report and Review of the Literature, 2017 DOI : 10.1177/1538574417702777,
22. Rong Xu Du , Bao Liu , Jiang Shao , Yu Chen , Fangda Li , Yuehong Zheng , Surgical Management of Congenital Arteriovenous Fistula near Lateral Cranial Base, 2017 PMID: 28687503 DOI: 10.1016/j.avsg.2017.06.123,
23. Ming Wang , Fan de Weijian , Rajneesh Mungur , Jun Gu , Shu Wan , Endovascular Treatment of Congenital Internal Carotid-Jugular Fistula, ,2018 PMCID: PMC6305339 DOI: 10.3389/fneur.2018.01118
24. Hu J, Zeng X, Yuan X. Fetal carotid-jugular fistula: A case report. Medicine (Baltimore). 2019 Jul;98(28):e16250. doi: 10.1097/MD.0000000000016250. PMID : 31305405; PMCID: PMC6641845.
25. Osborne PR, Sverzut TFV, Trivellato AE, Matias CM, Sverzut CE, Sverzut AT. Carotid jugular fistula after Le Fort I osteotomy. Int J Oral Maxillofac Surg 2017;46:845—50.
26. Guneyli S, Cinar C, Bozkaya H, Korkmaz M, Oran I. Endovascular management of congenital arteriovenous fistulae in the neck.DiagnInterv Imaging 2016;97:871.
27. Erdi F, Kaya B, Keskin F, et al. Congenital carotid–jugular fistula in a child: a case report. VascEndovasc Surg 2014;48:256–8.
28. Guneyli S, Cinar C, Bozkaya H, et al. Endovascular management of congenital arteriovenous fistulae in the neck. Diagn Interv Imaging 2016;97:871–5.