CLINICAL CASE OF SIMULTANEOUS RADICAL TREATMENT OF TETRALOGY OF FALLOT WITH MAJOR AORTOPULMONARY COLLATERAL ARTERIES

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The authors declare that they have no conflicts of interest

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Abstract

Tetralogy of Fallot (TOF) combined with major aortopulmonary collateral arteries (MAPCA) is a severe congenital heart defect due to the combination of a triad of cardiac malformation withan additional vascular anomaly of the small circulatory system. To date, there is no single accepted standard in which sequence and according to which criteria radical surgical correction of combined anomalies is indicated. This paper describes a clinical case of simultaneous opentreatment of TOF and endovascular occlusion of the MAPCA in an 8-month old child. Based on the evidence base of a large study, our patient belonged to the group where after TOF correction the therapeutic way of MAPCA treatment was used at first, which was ineffective in 38% and led to surgical methods of MAPCA occlusion. Choosing the way of simultaneous simultaneous surgical treatment of two pathologies allowed us to minimize postoperative risks and achieve good clinical results.

Үлкен қолқа-өкпе коллатералдары бар Фалло тетрадасын симуляцияланған радикалды емдеуді жүргізудің клиникалық жағдайы

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Аңдатпа

Фалло тетрадасы (ФТ) үлкен қолқа-өкпе коллатералдарымен біріктірілген жүрек ақауының триадасының кіші қанайналымы шеңберінің қосымша тамырлы ауытқуымен үйлесуіне байланысты ауыр туа біткен жүрек ақауы болып табылады. Бүгінгі күні біріктірілген ауытқуларды радикалды хирургиялық түзету қандай реттілікпен және қандай критерийлер бойынша орындалатыны туралы көрсетілген бірыңғай қабылданған стандарт жоқ. Бұл жұмыста 8 айлық балаға ФТ және эндоваскулярлық сәулелік окклюзияны ашық әдіспен бір мезгілде емдеудің клиникалық жағдайы сипатталған. Үлкен зерттеудің дәлелді базасына сүйене отырып, біздің науқас ФТ түзетілгеннен кейін алдымен сәулелерді емдеудің терапиялық жолы қолданылған топқа кірді, оның 38%-ы тиімсіз болып шықты, бұл сәулелерді окклюзиялаудың хирургиялық әдістеріне жүгінуге әкелді. Бірден екі патологияны симуляциялық хирургиялық емдеу жолын таңдау бізге операциядан кейінгі қауіп-қатерлерді азайтуға және оңтайлы клиникалық нәтижеге қолжеткізуге мүмкіндік берді.

Мүдделер қақтығысы
Авторлар мүдделер қақтығысының жоқтығын мәлімдейді

Түйін сөздер:
Фалло тетрадасы, үлкен аорто-өкпе коллатералдары, клиникалық жағдай, туа біткен жүрек ақауы, жүрек операциясы.

Клинический случай проведения симультанного радикального лечения тетрады Фалло с большими аорто-лёгочными коллатералями

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Ключевые слова:
тетрада Фалло, большие аорто-лёгочные коллатерали, клинический случай, врожденный порок сердца, операция на сердце

Аннотация

Тетрада Фалло (ТФ) в сочетании с большими аорто-легочными коллатералями (МАРСА) представляет собой тяжелый врожденным порок сердца, обусловленный сочетанием триады кардиального порока с дополнительной сосудистой аномалией малого круга кровообращения. На сегодняшний день не существует единого принятого стандарта в какой последовательности и по каким критериям показана радикальная хирургическая коррекция сочетанных аномалий. В настоящей работе представлено описание клинического случая одновременного лечения открытым способом ТФ и эндоваскулярной окклюзии БАЛК ребёнку 8 месяцев. Основываясь на доказательной базе крупного исследования, наш пациент входил в группу где после коррекции ТФ сначала применялся терапевтический путь лечения БАЛК, в 38% который оказывался неэффективным что приводило прибегать к хирургическим методам окклюзии БАЛК. Выбрав путь симультанного хирургического лечения сразу двух патологий, позволило нам минимизировать послеоперационные риски и добиться хорошего клинического результата.

Introduction

Tetralogy of Fallot (TOF) combined with major aortopulmonary collateral arteries (MAPCA) is a severe congenital heart disease aggravated by additional volume of cross over blood flow from the large to small circulatory system. There is no special standard for transcatheter occlusion of collaterals, there are no clear algorithms of surgical tactics, and therefore the surgeon faces the task of determining the indications and times tages of correction of combined TOF and MAPCA pathology. To determine the hemodynamic significance of MAPCA, and thus indicate the need for their one-stage closure, an attempt was made in The Second Xiangya Hospital of China on 380 patients operated on over a 10-year period [1]. These patients accounted for 28.1% of all operated TOF (1351 cases), of which MAPCA occlusion was performed in 4.8% of cases. A mathematical formula for the MAPCA occlusion index was derived, where $K=((\sum R^2)/Wt)$, R is MAPCA diameter and Wt is patient weight. With $K\geq2$, collateral occlusion and surgical correction are recommended to be performed simultaneously, with $1< K< 2$, the need forcollateral occlusion depends on the postoperative condition of patients with more than the standard increase in artificial pulmonary ventilation (APV) time for this pathology. In this group, the need for MAPCA occlusion was 38%.

During radical TOF correction, if hemodynamically significant MAPCA are not closed, severe congestion in the small circle of the circulation and bypass of the great circle of thecirculation are expected in the postoperative period [2]. In such situations, a longer period of ventilatory ventilation is required and many patients have complications such as low cardiac output syndrome, pulmonary edema, lung infection and pleural effusion. The tandem approach, in which endovascular MAPCA occlusion immediately after radical open surgery is the second stage, effectively solves this problem [3].

Case study

A child 8 months old, 7 kg was admitted to the clinic on 09/28/2022 with diagnosis: TOF, PFO, PDA, MAPCA with complaints of shortness of breath, lividity of the skin, rapid fatigue during feeding. The basic diagnostic tests: ECG, echocardiography (ECHO) were performed. Instead of contrast computed tomography (CT), cardiac catheterization (CC) was performed. The defect was confirmed by ECHO data: Aorta: 1.4 cm, bulbus shifted to the right by 50%. MAPCA of 0.3 cm was not excluded. Pulmonary artery (PA): valve 0.55 cm; flaps compacted, trunk 0.6 cm. The branches of the pulmonary artery are 0.6 cm each. The gradient on the PA valve was 76/45 mmHg. Degree 0-1 regurgitation. Interventricular septum: subaortic defect 1.1 cm. Interatrial septum: PFO 0.15cm. Cardiac cavities: right sections moderately dilated, Myocardial contractility: satisfactory. To clarify cardiac anatomy and extracardiac vascular structures, the following was performed: CC from the right ventricle (RV)- the aorticarch with the PA trunk and branches was contrasted simultaneously, the RV cavity increased in volume, moderate hypoplasia of the PA trunk and valve ring, moderate kinking of the left PA branch mouth, the middle and distal segments of PA branches were developed satisfactorily (Fig. 1). There was marked tubular stenosis in the exiting RV. In thoracic aortography, there is contrasting hypervascularization zones of both lungs across all fields, due to the presence of a developed MAPCA - bronchial and intercostal arteries branching off in the projection Th 2-3 of the thoracic spine, with adiameter of MAPCA afferent arteries of 1.5-3.0 mm (Fig. 2). Tonometry in mm Hg: PA trunk 30/5, middle 13, PJ75/3, distal segment of right and left branches of PA 24/7 (middle 12). The diameter of the distal segments of the PA right 6.4 mm (Z score 0.03), left 7.6 mm (Z score1.69) PA trunk 7.3mm (Z score-2.12) (Fig.3), PA valvering 5.5 mm, aorta valvering 13.8 mm, at diaphragm level 6.1 mm.

Diagnosis

TOF, subclavian stenosis with moderate hypoplasia of the valvering and PA trunk. Developed MAPCA network on both sides.

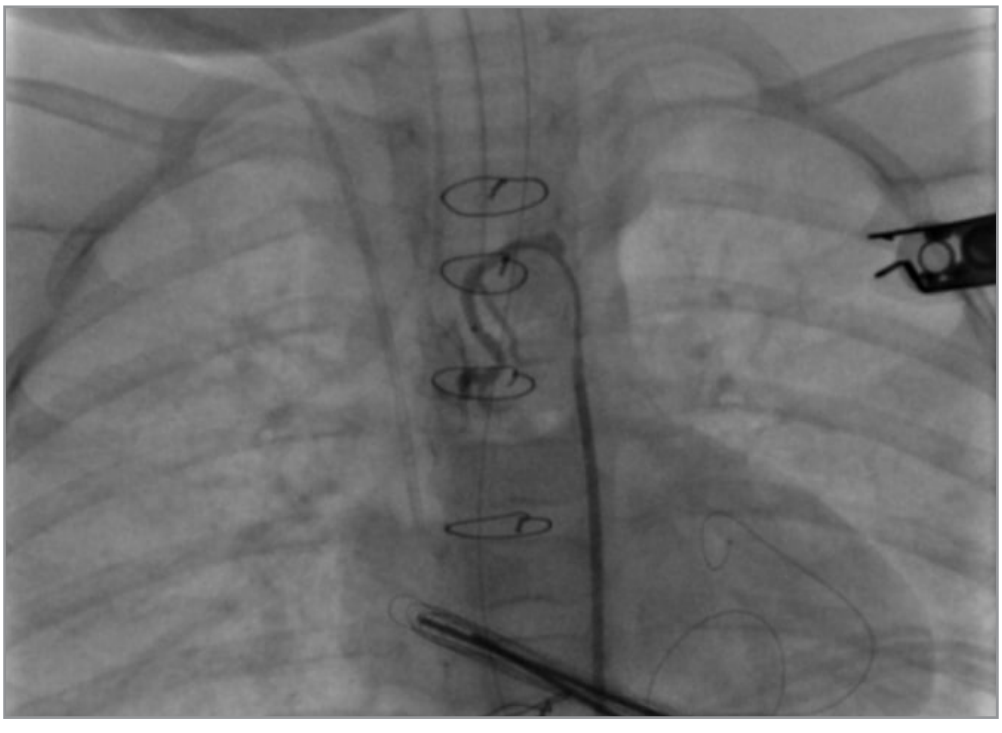


Figure 1.
Angiogram. Enlarged and hypertrophied right ventricle, exit section with marked tubular stenosis, hypoplasia of the pulmonary artery trunk and valvering, satisfactorily developed pulmonary artery branches

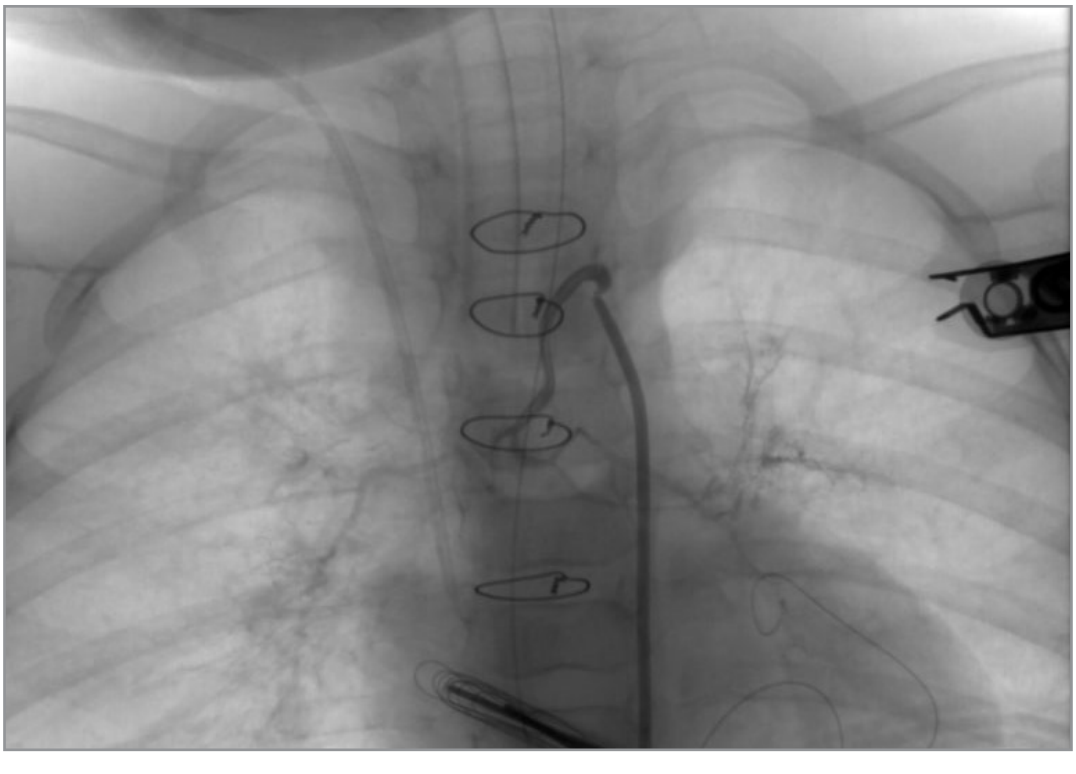


Figure 2.
Angiogram. MAPC - bronchial branch originating from the arch B of the aortic segment with a diameter of 3.0 mm

Figure 3.
Z score of the pulmonary artery branches and trunk by cardiac cavity probing

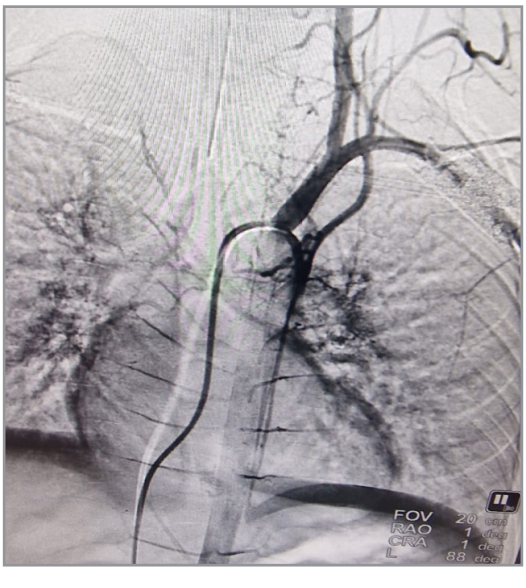
Height (cm): 68
Weight (kg): 7
BSA formula: DuBois 0.35 M^2

Site	Measured (cm)	Mean	Range	Z-Score
RVD:		1.21	(0.81 - 1.80)	
IVSd:		0.41	(0.29 - 0.59)	
IVSs:		0.57	(0.42 - 0.77)	
LVIDd:		2.39	(2.02 - 2.81)	
LVIDs:		1.48	(1.20 - 1.82)	
LVPWd:		0.34	(0.25 - 0.47)	
LVPWs:		0.66	(0.51 - 0.84)	
Aortic Annulus:		0.88	(0.74 - 1.03)	
Sinuses:		1.22	(1.02 - 1.46)	
ST Junction:		0.97	(0.77 - 1.20)	
Transverse Arch:		0.99	(0.77 - 1.28)	
Isthmus:		0.69	(0.53 - 0.91)	
Distal Arch:		0.74	(0.56 - 0.96)	
Ao at Diaphragm:		0.71	(0.57 - 0.88)	
Pulmonary Annulus:	0,73	1.01	(0.78 - 1.29)	-2.12
MPA:		1.05	(0.81 - 1.36)	
RPA:	0,64	0.64	(0.49 - 0.84)	0.03
LPA:	0,76	0.57	(0.43 - 0.75)	1.69
Mitral Annulus:		1.48	(1.16 - 1.90)	
Tricuspid Annulus:		1.61	(1.18 - 2.20)	
Left Atrium:		1.45	(1.15 - 1.83)	

Update reset

On 11.10.2022 we performed elective surgical correction under the conditions of cardiopulmonary bypass: VSD repair with anautopericardial patch, in fundibulotomy, PA valve commissurotomy with valve and PA fibrous ring preservation, RV outlet plasty with anautopericardial patch, PFO suturing. After the operation the child was immediately taken from the cardiac surgery room to the Cath-Lab operating room. Course of intervention: retrograde puncture and catheterization of the right common femoral artery by Seldinger. A JR 3.5 4Fr type catheter was and placed on the aortic arch, and thoracic aortography was performed, in which contrasting zones of hypervascularization of both lungs were noted throughout the fields, due to the presence of a developed 3 mm diameter MAPCA system (Figure 4). The source of MAPCA was a bronchial branch originating from the aorticarch. Given the angiographic findings, MAPCA embolization was recommended. A BS Renegade microcatheter was installed at the kostium of the pathological branch, and embolization with BS Contour 45-150μM pVa particles - 1 vial was performed. At the control angiography the contrast intensity was reduced by 100% (Fig. 5). There were no complications during the operation.

Figure 4.
Angiogram. Catheter inserted into MAPCA, thoracic aortography with contrasting areas of hypervascularization of both lungs across all fields



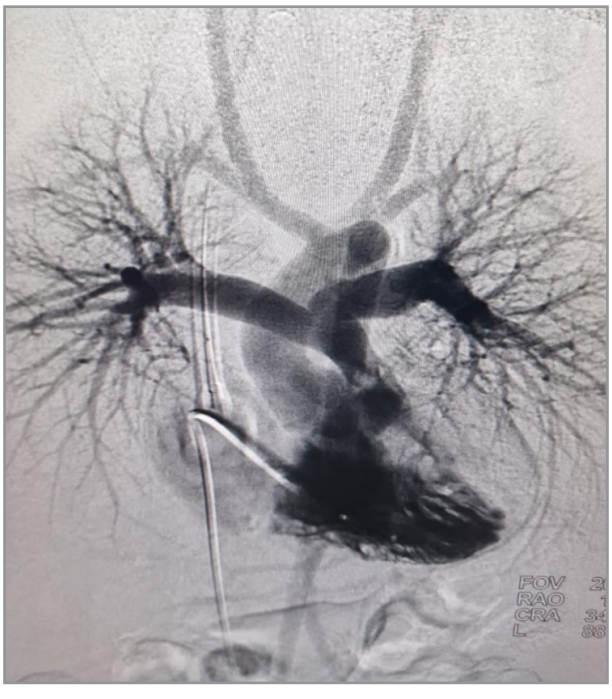


Figure 5.
Angiogram. End result after MAPCA embolization: at control angiography, the contrast intensity of pulmonary vascularization was reduced by 100%

Control postoperative ECHO: tricuspid valve 1st degree regurgitation. Mitral valve: 0-1 degree regurgitation. Aorta: 0-1 degree regurgitation. Abdominal aorta: main blood flow. Pressure gradient on the PA valve: 10/5 mmHg, regurgitation 0-1 degree. Left ventricular ejection fraction 64%. Interventricular septum: patch, shunt 1 mm. Atrial septum: tightly sutured. The child was extubated 44 hours after the simultaneous surgeries, on the 6th postoperative day he was transferred to the specialized department and discharged on the10th day.

Discussion

MAPCA is one of the significant factors influencing the final outcome of radical TOF correction [4]. At the early stage, cardiac surgeons did not know much about collaterals and did not treat them when performing radical TOF correction. With the development of medical and computed tomography technologies, cardiac surgeons gradually deepened their knowledge of aortopulmonary collaterals and their treatment became an important part of radical TOF correction [5].

It is logical to try to close all available additional collaterals in open surgery in one step after cardiac TOF correction, but technical difficulties in the form of collateral search, expansion of the operating field and surgery time, in most cases the inability to close all sources of additional blood supply, have led to the development of a staged approach of radical treatment involving interventional technologies. Endovascular techniques have a number of advantages over hard-to-reach surgical areas and are used both before, during, and after open surgery [6].

At the current stage, when the task is to perform simultaneous cardiovascular surgical interventions, a universal hybrid operating room [7], which allows performing open and closed surgery simultaneously, is deployed on the basis of highly specialized clinics. A tour center, we were able to minimize the time factor between the first and second stages of TOF radical correction with MAPCA, thanks to the functional location of two closely situated multi-targetoperating theatres on one floor of the surgical unit. Most studies on MAPCA treatment have focused on MAPCA occlusion before TOF correction, but MAPCA occlusion before surgical correction could lead to a further decrease in saturation, and therefore the patient required surgical correction immediately after transcatheter closure of MAPCA.

To reduce morbidity and mortality, endovascular MAPCA occlusion or surgical ligation should be performed in patients with a collateral diameter to body weight ratio of at least 0.5 mm/kg. In patients with values of approximately 0.2 to 0.5mm/kg, prolongation of APV should have priority over transcatheter occlusion, and for patients with values below 0.2 mm/kg no additional treatment is required [8]. In our child's case, this value left 3.5 in the presence of 3.0 mm collaterals. We prioritized the closure of collaterals immediately after TOF correction with the desire not to prolong APV, which would have increased the risks of APV-associated complications.

Conclusion

In our clinical case based on the experience of the world leading clinics, the principle of simultaneous treatment of severe combined pathology of Tetralogy of Fallot with major aortopulmonary collateral arteries was successfully applied. Taking into account the evidence base, our patient belonged to the intermediate group, where single-stage correction of TOF with MAPCA occlusion was performed in 38% of cases. During radical open correction of TOF, the valve complex of the PA trunk was preserved and MAPCA was embolized immediately within several minutes after the first operation, which allowed to minimize postoperative risks and achieve a good clinical result.

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