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Review Article

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Role of Brown Adipose Tissues / Beige Adipose Tissue (BAT/bAT) relevant genes expression and polymorphism in regulating human body weight

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Summary

Introduction: *Tetralogy of Fallot is the most common cyanogenic congenital heart disease. In sub-Saharan Africa, the incidence of the disease is unknown. Hospital studies show that it is the most common cyanogenic congenital heart disease. This heart disease, which varies in severity, has benefited from considerable medical progress over the last fifty years. The main objective of the work is to study management modalities of this heart disease in Senegal.*

Method: *This retrospective study conducted from first January 2010 to thirty-one December 2015 in the pediatric cardiology department of the Albert Royer National Children Hospital of Dakar (CHNEAR), involves 125 children diagnosed with tetralogy of Fallot.*

Results: *Tetralogy of Fallot accounted for 18.6% of congenital heart disease. Inbreeding and trisomy 21 were the main risk factors. Cyanosis was the main sign of disease discovery (33.2%) followed by anoxic malaise. Pulsed oxygen saturation averaged 70%. Severe acute malnutrition was detected in 21.8% of our patients. Ultrasound showed a regular pattern in 81 patients (64.8%) and an irregular pattern in 44 cases (35.2%). Modified Blalock surgery was performed in 16 patients (12.8%) and a surgical cure in 43 patients (34.4%). We reported 20 (16%) deaths in this series.*

Conclusion: *The results of our series show an improvement in the management of Fallot tetralogy in Senegal, but the data is insufficient. Effort must be made in the context of disease screening and early surgical management.*

Keywords: Tetralogy of Fallot; Cyanosis; Blalock Surgery; Senegal

Introduction

Tetralogy of Fallot is the most common cyanogenic congenital heart disease with an incidence of approximately 0.5/1000 live births [1-10]. It statistically accounts for 5-8% of congenital heart disease [4, 10]. BARAKAT found in one of his hospitals studies an incidence of 4.9/1000 live births. Management of this curable heart disease requires a specialized environment, in particular an operating room adapted for pediatric cardiac surgery as well as a pediatric and neonatal intensive care unit [1]. Currently, Fallot tetralogy screening is performed at a young age and even antenatally in developed countries. In sub-Saharan Africa, particularly in Senegal, despite many advances in the management of Fallot tetralogy, predicaments in its management still prevail.

This delay in management leads to long-term complications of "aged Fallot". The new medical-surgical cardio pediatric center CUOMO inaugurated in Dakar in early 2017, raises hope for surgical management of children with heart disease in the country.

Methodology

This is a retrospective study of 125 cases of tetralogy of Fallot collected in the cardiology department of CHNEAR. We included all children with a tetralogy of Fallot confirmed by cardiac ultrasound followed in the cardiology department of the Albert Royer Children's Hospital in Dakar from January first, 2010, to December thirty-one, 2015.

Results

During the study period, 672 children with congenital heart disease were included in CHNEAR's cardiology department. Tetralogy of Fallot accounted for 18.6% of congenital heart disease. The sex ratio is 1.6. Inbreeding was noted in 38.9% of patients. Symptoms began on average at 8.6 months [range 0-36 months]. Age at diagnosis ranged from birth to 120 months with a mean of 20.19 months. The time from symptom onset to age at diagnosis averaged 11.5 months. Cyanosis was the main sign of disease onset (33.2%) followed by dyspnea (31.1%), anoxic discomfort (12.8%), and squatting (10.1%). (Table 1) illustrates the circumstances of the discovery of Fallot's tetralogy and (Figure 1) the main physical signs.

Table 1: Circumstances of Discovery of the Tetralogy of Fallot

Circumstances of Discovery	Case	Percentage (%)
Dyspnea + fatigability	30	24
Cyanosis	36	28,8
Squatting + walking fatigue	17	13,6
Delayed psychomotor development	1	0,8
Anoxic discomfort	29	23,2
Malnutrition/Staturo-ponderal Retardation	1	0,8
Malformative assessment	2	1,6
Heart murmur + cyanosis	2	1,6
Preoperative assessment	2	1,6
Sepsis	2	1.6
Repeated lung infections	3	2,4

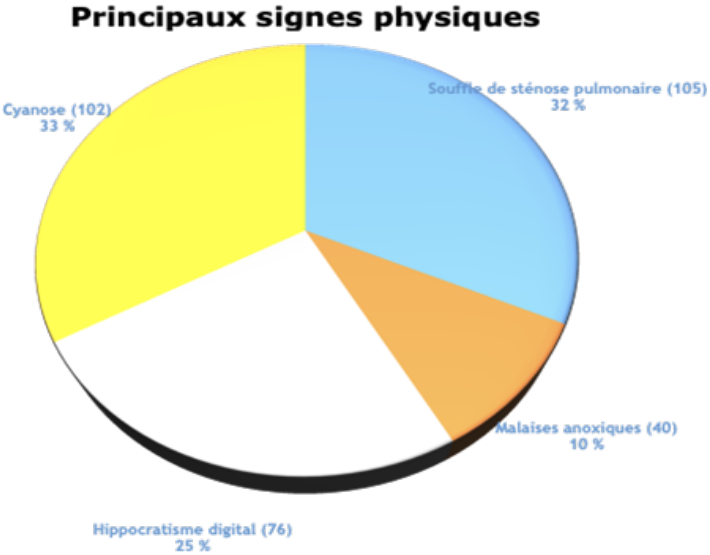

Figure 1: Main physical signs of Fallot's tetralogy

Nutritionally, 17 patients (21.8%) were severely acutely malnourished with a weight-for-height ratio [-3; -4 Z-score] and 75 patients (60%) were underweight with a weight-for-age ratio < -3 Z-score. Isolated forms of Fallot tetralogy were the most common: 112 patients (89.6%). A genetic syndrome or associated malformation was noted in 13 patients (10.4%). None of the patients benefited from a cytogenetic study. We found 8 cases of trisomy 21 (6.4%); 2 cases of omphalocele (1.6%); 2 cases of VACTERL syndrome (1.6%); 1 highly suspicious case of trisomy 18 (0.8%). Polyglobulia was noted in 41 patients (38.7%); Anemia was present in 12 patients (11.3%). All patients showed electrical signs of right ventricular hypertrophy (RVH) on ECG; the electrical axis of the heart averaged +135°. Chest X-ray showed a 'heart in the hoof' in 73.4% of patients, cardiomegaly (TBI > 0.55) in 80.5% of patients, and pulmonary hypo-vascularization in 61.9%. Trans-thoracic ultrasound showed a regular shape in 81 patients (64.8%) and irregular shape in 44 cases (35.2%). Pulmonary stenosis was essentially infundibular (60.8%); the max gradient Right Ventricle - Pulmonary Artery (mmHg) averaged 82.4 mmHg. The pulmonary pathway was normal in 72 patients (57.6%). There were 14 congenital heart diseases associated with Fallot tetralogy (11.2%). There were: 6 atrioventricular channels (4.8%), 6 cases of persistent ductus arteriosus (4.8%); 2 atrial septal defects (1.6%). Therapeutically, 113 patients (90.4%). were on beta-blockers. Iron supplementation was indicated in 51 patients (40.8%). Surgically, 16 patients (12.8%) received modified Blalock anastomosis. The time from diagnosis to modified Blalock anastomosis averaged 15.4 months. The mean age at the time of modified Blalock surgery was 28.6 months. A complete cure was recorded in 43 patients (34.4%). In addition, it should be noted that 25 patients (58.1%) were operated in Europe, while 18 underwent surgery (41.9%) in Dakar. The mean age at the time of surgical treatment was 103 months and the time between diagnosis and surgical treatment averaged 41 months.

Complications were detected in 20 patients (16%). Infectious complications occurred in 14 patients (11.2%), with mainly cerebral abscesses (4.8%) followed by recurrent pneumonia (3.2%). Vascular complications were found in 6 patients (4.8%) with mainly cerebral thrombosis (3.2%). 18 patients (14.4%) who did not receive surgical management, deceased.

The overall postoperative outcome following a complete surgical treatment was favorable/acceptable. However, some complications were noted with one case of conduction disorders (complete AVB); 4 cases of residual pulmonary stenosis, and mild pulmonary insufficiency in 3 patients. There were 2 postoperative deaths due to hemorrhagic syndrome increasing the number of deaths to 20 (16%).

Discussion

The prevalence of Fallot tetralogy in our series was 18.6%. Our results are consistent with several African studies. In fact, KAKOU found a prevalence of 15.5%, BARAKAT 16.6%, and DIOP 16.49% [2, 6]. The age at diagnosis is 20.19 months, in line with some African studies. (Table 2) compare the age at diagnosis of Fallot's tetralogy in different studies and the (Table 3) compare the summary of clinical signs of Fallot's tetralogy between different series. Results discrepancy with Western studies is justified by the early diagnosis of Fallot tetralogy, generally, in antenatal care. In comparison, DIOPs series completed from 1992 and 1995 in

Senegal revealed an earlier age at the time of diagnosis. This finding indicates an improvement in the means of diagnosing congenital heart disease in Senegal over time. In our study, 64.8% regular and 35.2% irregular shapes were identified. The ELYANDOUZI series also conceded the regular form to be the most frequent with 67% of the cases; (78.46%) in the LAMLIKI study and 57.7% in KAKOU study. A brain abscess was noted in 4.8% of our patients; whereas it was lower in the DIOP series (2.9%); and non-existent in the ELYANDOUZI and LAMLIKI series. In our palliative series, Blalock-modified surgery was performed in 12.8% of patients. Curative surgery was executed in 30.4% of patients. In the DIOP series undergone years ago (1992-1995), only 20.6% of patients were able to benefit from curative surgery. The mean age at the time of Blalock-modified surgery was 28.6 months in our series, which was greater than in the ELYANDOUZI series of 18 months; but lower than in the KAKOU series of 56 months and in the THIAM series of 57.6 months. The mean age at the time of curative surgery was 103 months; while in the ELYANDOUZI series, it was 58 months. The age was greater in the KAKOU series where it was 91 months. In the THIAM series, the average age at the complete cure was 83.35 months. Results found in the sub-Saharan African series, particularly in ours, testify to the adverse effects in the late treatment of Fallot tetralogy.

Table 2: Age at diagnosis of Fallot's tetralogy in different studies

Author	Location of the study	The average age of diagnosis (months)
GUIRGIS [5]	France	3,7
NEED [8]	UNITED STATES	4,9
ELYANDOUZI [3]	Fez (Morocco)	30
LAMLIKI [7]	Rabat (Morocco)	48
BARAKAT	Lagos (Nigeria)	51,6
KAKOU	Abidjan (Ivory Coast)	136,8
DIOP	Dakar (Senegal)	100,3
Our series	Dakar (Senegal)	20,19

Table 3: Summary of clinical signs of Fallot's tetralogy between different series

Clinical events	Our study (%)	ELYANDOUZI (%)	LAMLIKI (%)	DIOP (%)	BARAKAT (%)	THIAM [9] (%)
Cyanosis	81,6	95	98,46	79,41	72,1	-
Dyspnea	81,6	85	40	94	4,2	79,4
Anoxic discomfort	32	43	36,9	17,6	-	44,1
Squatting	32	31	21,5	76,47	2,4	55,9
SaO2	70	74	-	-	91	-
Digital Hippocratism	60,8	38	30,8	70,58	-	-
Delayed psychomotor development	4,6	-	3,02	-	-	-
Staturo-ponderal delay	20,5	5	26,15	-	0,6	5,9
Breath of pulmonary stenosis	84	99	-	-	-	23,5

Limitations of our study

The limitations of our study are mainly related to its retrospective nature. We had difficulties in exploring our files, while occasionally missing complementary examinations.

Conclusion

Even though the results of our series indicate an improvement in the management of Fallot tetralogy in Senegal, this is still insufficient. These results are encouraging, given that patients with Fallot tetralogy did not previously receive adequate care. A great deal of effort must be made in the early detection of this cardiopathy. The new medical-surgical cardiopediatric center CUOMO inaugurated in Dakar in early 2017 raises hopes for the surgical management of children with heart disease in Senegal.

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