ANOMALOUS RIGHT CORONARY ARTERY CONNECTION TO THE OPPOSITE SINUS WITH AN INTER-ARTERIAL-PULMONARY COURSE: DIRECT REIMPLANTATION TECHNIQUE WITHOUT TRANSVERSE AORTOMY.

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Abstract

Introduction:

The anomalous connection of the right coronary artery to the left posterior aortic sinus associates an inter-arterial-pulmonary course, an intramural course and ostial anomalies.

They are known to be a cause of sudden death in athletes. There is no recommendation of treatment strategy. The surgical treatment in is very controversial, especially in adults.

Material and method:

This is a retrospective unicenter study of 30 cases of patients who received surgery operated in our center between January, 2003 and December, 2016.

Results:

16 patients were less than 18 years old and 10 over 30 years old. 14 patients practiced sport.

86, 6 % were symptomatic with 5 sudden deaths.

The average time between diagnosis and intervention was 18, 5 months (0, 5-179). All the patients had a cardiac CT scan and a reimplantation in the anatomical position.

The average follow-up was of 25, 6 months (1-701) with a death after 3 post-operative months. At the time of the latest news , 92, 8% were NYHA I and 60 % had returned to a physical activity.

Conclusion:

Only the reimplantation allows to restore the coronary anatomy and enables patients to return to a physical activity.

The evolution or the long-term appearance of a coronaropathy on this right reimplanted coronary artery must be assessed.

1.Introduction :

The abnormal connection of a coronary artery to the opposite sinus (AACCA) may concern the left coronary artery (Anomalous Left Coronary Artery, ALCA) as well as the right coronary artery (Anomalous Right Coronary Artery, ARCA). The ARCA may then be associated to an anomalous course(1) between the aorta and the pulmonary artery (Inter-arterial Communication, IAC) from the right ventricular auricle channel. It is considered that 0.1% to 0.3% of the population is concerned by these anomalies. The exact prevalence of AACCA in the general population remains difficult to determine and has been for a long time estimated using a case series or autopsy series (2).The AACCA are known to be the second cause of death by sudden death in athletes (3). The improvement of the screening techniques and imaging techniques has substantially increased the prevalence of these anomalies these past ten years (4).

However there is no recommendation on treatment strategy and systematic surgical management remains a topic of debate. Several surgical techniques have been described.The the absolute necessity of the surgical management of the . ALCA with IAC is well known, the one of ARCA with IAC particularly in adults is very controversial(5). We describe a series of 30 patients who received surgery in our center between January 2003 and december 2016 by direct reimplantation for an ARCA with IAC, of which 27 received surgery without transverse aortotomy.

2.Patients and methods :

We created a cohort of patients from our database who underwent surgery for an ARCA with IAC with a technique of direct reimplantation between January 2003 and December

2010 .Our study is a unicenter retrospective study. This is a case study. We have created a file of anonymous records and obtained the approval of the clinical investigation center. The follow-up was done in the immediate postoperative period and at the date of the latest news on the data of the consultation reports.The clinical improvement of the survivors has been assessed on the presence or not of symptom, the return to a physical activity and the absence of ischemia signs on the electrocardiogram or on the Holter ECG.The coronary reimplantation was assessed by two-dimensional transthoracic echocardiography with doppler and a cardiac CT scanner and/or coronary angiography .The left ventricular function was assessed by two-dimensional transthoracic echocardiography on the shortening fraction. The evolution of the outbreak of a postoperative myocardial ischemia was assessed either by a stress test or by a thallium myocardial scintigraphy.

2.1. Statistics

Mean median and standard deviation calculation with a confidence interval of 0.25.Nonparametric Wolcoxon tests were performed on matched series for means comparison. Fisher tests performing for proportion comparisons when the effects were inferior to 5. Significant if p-value less than <0.05.

2.2 Population

Between January 2003 and December 2016,30 patients have received a surgery for an ARCA with IAC.Amongst these 30 patients,16 were less than 18 years old and 10 were over 30 years old at the time of the surgical procedure (0.18-52) (Figure 1).

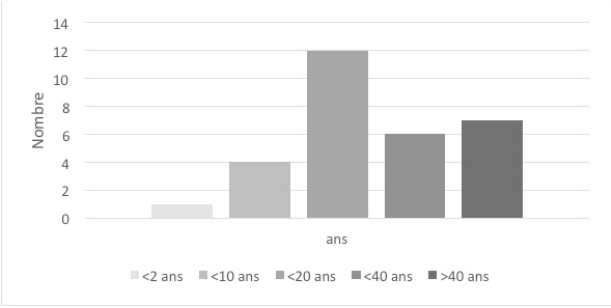

Figure 1. Age distribution at the time of surgery (number/year) of patients who underwent a surgery for ARCA (Anomalous Right Coronary Artery) with IAC (Inter-Arterial Communication)

The m/f sex ratio was 4. The mean weight at the time of the surgery was 58 Kilograms (3,6-118).20% of the patients had at least one cardiovascular risk factor.2 patients were

active smokers,3 were overweight (BMI>27),1 had a chronic AHT in the context of a primary hyperaldosteronism,1 had an essential AHT and one had a non-insulin-dependant diabetes. No family history of coronary anomaly was found but the mother of a patient died from Uhl Anomaly.5 patients(16,6%) had an associated chronic pathology (1 had Kawasaki disease without coronary heart disease,1 had epilepsy,1 ankylosing spondylarthritis,1 Fabry disease,1 Guillain-Barré syndrome. No patient was syndromic.7 patients had an associated cardiac malformation (1 had a coarctation of the aorta, 1 a perimembranous ,ventricular Septal Defect (VSD),1 pulmonary atresia without VSD (with residual IAC), 2 had Interatrial Communication (IAC),1 had biscupid aortic valve disease and 1 had left ventricular non compaction (LVNC).14 patients (46,6%) were regularly practicing sports,and 7 of them in competition (23,3%).

2.3 Clinical presentation

86,6% of the patients were symptomatic with 5 patients(16,6%) having recovered from sudden death.1 of these patients only had prodromes and a regular cardiovascular follow-up before the sudden death episode.6 patients have had 1 or more episodes of syncopal episodes (20%), 20 had chest pains (66%).
In 63% of cases the pains are recorded during or immediately after an effort.
For 3 patients,an infectious context associated with atypical chest pains lead to a cardiological check-up enabling the diagnosis. .2 patients have been diagnosed following up an examination for heart murmur and 1 during a follow-up of Fabry disease.1 ARCA was discovered following a check-up for attacks of acute Cardiac Arrhythmia/poorly tolerated Atrial Fibrillations (Table 1).

Table 1. Clinical and postoperatory presentation of the patients.

	Sex	Age at time of surgery (years)	Clinical signs	Sport (1=yes 0=no)	Risk factors	Associated malformation	Period before surgical procedure (months)	Associated gesture	Postoperative NYHA	LFU
1	M	17	chest pain	1			4		I	
2	M	52	chest pain	0			6		I	
3	M	11	chest pain	1			107		I	
4	M	32	chest pain	0	tobacco		1		I	chest pain
5	F	51	chest pain	0			1		II	chest pain
6	M	48	passage CAr/AF	0		Bicuspid Aortic Valve	28	AVR	I	
7	M	10	syncope chest pain	1			14		I	
8	M	11	chest pain	1			2		I	
9	F	40	chest pain	1			3		I	
10	M	10	chest pain	0		VSD operated	19		I	
11	M	23	chest pain	0			9		I	
12	M	16	chest pain	0			3		I	
13	M	9	0	0			5		I	
14	M	5	heart murmur	0			35		I	
15	F	52	chest pain	0	NIDDM		5		II	
16	F	7	syncope chest pain	1		Coarctation operated	43		I	
17	M	0,18	sudden death Fetal Arrhythmia	0		FOP	2	FOP	II	DEC
18	M	24	chest pain	1	AHT		3		I	
19	M	23	sudden death	1	AHT	LV Non compaction	0,5		I	
20	M	17	sudden death	0			1		I	
21	M	17	chest pain	1		IAC	5	IAC	LTFU	LTFU
22	M	15	chest pain	0		PAIVS residual IAC	179	IAC PVR	II	chest pain
23	F	17	sudden death prodromes	0			3		I	Syncope defibrillator
24	M	19	heart murmur	1			2		I	
25	M	46	chest pain	1			2		I	chest pain
26	M	14	syncope chest pain	1			13		I	
27	M	7	syncope chest pain	1			10		I	
28	M	34	chest pain	1	tobacco		2		I	
29	F	51	syncope chest pain	0			46		I	
30	M	38	syncope chest pain	0			2			

Ao: aortique, AHT: Arterial Hypertension, VSD: Ventricular Septal Defect, FOP: Foramen Oval Permeable, IAC Interatrial Communication
LV Left Ventricular, DEC: deceased, LTFU: Lost to follow up
PVR:Pulmonary Valve Replacement , PAIVS Pulmonary Atresia with Intact Ventricular Septum ,CAr/AF Cardiac Arrhythmias /Atrial Fibrillation
AVR Aortic Valve Replacement NIDDM : Non-Insulin-Dependant Diabetes Mellitus
DDN :Last Follow Up

2.4 Preoperative check-up:

All the patients underwent a ECG, a frontal chest X-ray,a two-dimensional transthoracic echocardiography with doppler and a cardiac CT scan (Figure 2).

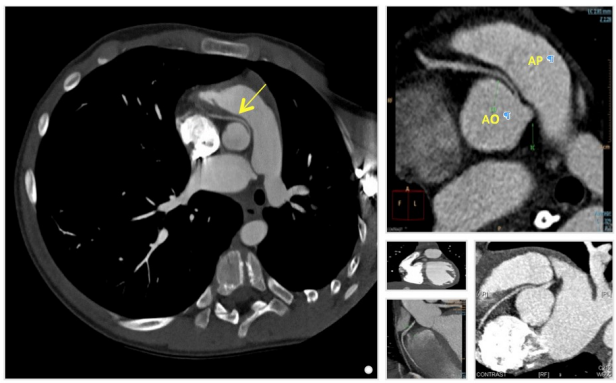

Figure 2: Serie of scan views of ARCA (arrow) with inter-aortopulmonary course in transverse and coronal sections.

At the ECG,3 patients had a sequelar Q wave and 7 a negative T wave in the inferior leads.5 patients had an incomplete right bundle branch.4 patients have had a non-contributory holter-ECG . The two-dimensional transthoracic echocardiography with doppler enabled to make the diagnosis in 83% of cases (Figure 3).

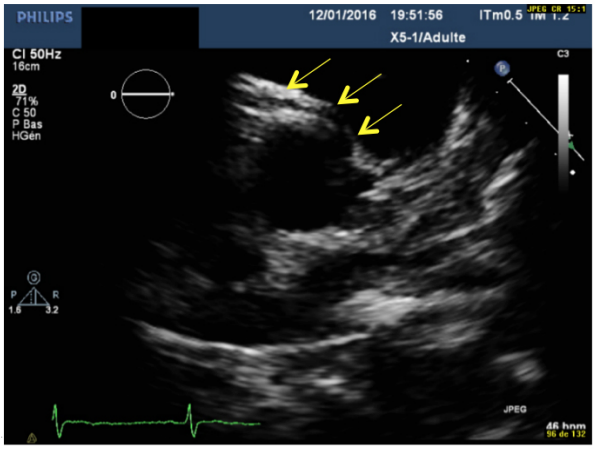

Figure 3. TTE showing the inter-aortopulmonary course of an ARCA

1 patient had a transesophageal echocardiography.The mean shortening portion was of 39%. 7 patients have had magnetic resonance imaging of which 2 for suspicion of myocarditis ,3 in order to eliminate a left ventricular arrhythmogenic dysplasia, and 1 enabling to make the diagnosis of a non compaction of the LV.

Likewise,7 patients,all adults,have had a coronary angiography.

14 patients underwent a stress test of which 4 of them electrically positive (3 ST-segment depression in V1,V2 and V3 and 1 sustained BES) and 2 clinically positive with painful relapse.5 patients had an exploration by thallium myocardial scintigraphy but only one has highlighted sequels of myocardial ischemia.10% of patients had a coronary heart disease, but not significant (stenosis < 50%) on the right or left network.

2.5 - Surgical technique:

The average period between the diagnosis and the surgical intervention was 18,5 months (0,5-179).The average age at the time of the surgical procedure was 24,2 years old (2,2 months old -52 years old).Since 2003,the number of patients referred to for surgery is increasing,with a maximum of 8 patients operated in 2015 (Figure 4).

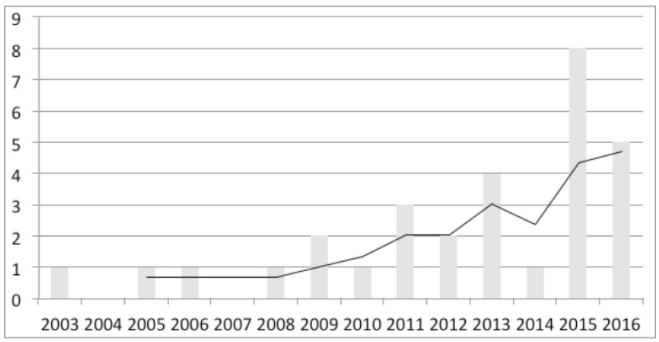

Figure 4. Distribution of patients operated per year.

Trend curve (based on 3 years average).

All the patients have been operated under ECC by median sternotomy.2 patients had already had a median sternotomy ,one of them had already had 2 median sternotomies (1 IAC closure,1 pulmonary valve replacement an then a systemic-pulmonary shunt) and a left thoracotomy patient for a coarctation treatment. These abnormalities had gone unnoticed. The mean time of ECC was 52 minutes (32-108) and the mean time of aortic clamping was 29 minutes(16-68).

There have been 5 associated gestures : 1 aortic valve replacement by a by a mechanical valve,1 pulmonary valve replacement by a bioprosthesic valve with an arterioplasty to enlarge the pulmonary artery trunk,and 3 closures of IAC (intra-auricular septal defects), (1 directly and 2 by patch) .All patients had a direct reimplantation of ARCA in the anatomical position.

All the reimplantations have been associated to an ostial plasty either by incision along the posterior wall or using a patch for enlargement (3 patients).

90% of the patients have had a direct reimplantation without transverse aortomy using a technique developed in our center.

2.5.1 Technique of direct reimplantation without transverse aortomy :(Figure 5)

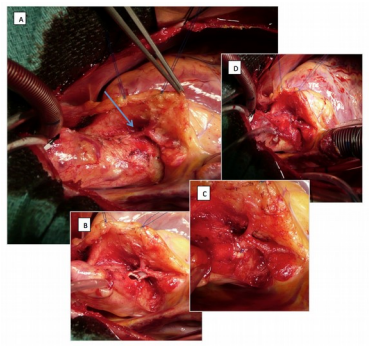

Figure 5. Technique of direct reimplantation in the anatomical position without transversal aortotomy.

A : mobilization of the right coronary artery and of the infundibular branch(arrow showing the ARCA) ;

B : aortotomy with 4 mm diameter incision ;

C : direct reimplantation in pallet by means of continous stiches ;

D : aorta of the right coronary after declamping in the anatomical position.

Under ECC at normothermia. Mobilization of the right coronary artery in the first centimeters before its connection to the aortic wall,on beating heart (A). Aortic clamping and undertaking of a anterograde cardioplegia in a crystalloid solution.Coronary artery transection at its connecting point to the aortic wall. Closure of the aortic entrance hole by means continuous stiches "back and forth".Performance of an circular aortotomy with incision (B).The coronary artery is cut "into appropriate lengths". Performance of a counterincision in its posterior side to enable a reimplantation "in pallet" (C).Reimplantation of the coronary artery on the aortic aorteriotomy by means of continuous stitches (D).

2.5.2 Direct reimplantation technique with transverse aortotomy

In equal operational conditions,the coronary is reimplanted in a direct way in the arterial wall after creating a "cubicle" and performing an anterior incision enabling an enlargement of the ostium using a autologous pericardium patch as described by Gaudin and al (10).

2.6 Postoperative result.

There was no postoperative complication. No patient had postoperative circulatory assistance such as ECMO.The mean time period before extubation was 5 hours (2-48),the average time spent In-Hospital Resuscitation was1,1 day (1-4).
30 days later,no postoperative deaths had occurred .There have been 5 complications: 2 pneumopathys treated by antibiotic therapy,1 pleural effusion having required drainage, 1 postoperative cardiac failure requiring an aminergic support longer than 2 days and a mediastinitis requiring a return to the operating room. The average hospitalisation time period was 7,5 days (5-20).

2.7 Follow-up :

The average follow-up period was 25,6 months (1-101),with 1 death at 3 months postoperatively and 1 was lost to follow up.
At the date of the latest news,26 patients were suffering from moderate heart failure NYHA Class 1,92,8 % of them.4 patients kept having chest pains,60% of patients had returned to a physical activity.16 patients were following a drug therapy of which 68,7% were taking Beta-Blockers. All the patients received aspirin therapy during 3 postoperative months.
3 patients kept a T negative wave at ECG and of the 8 patients having had a holter ECG, no conduction or rhythm trouble was highlighted.A 2 dimensional transthoracic echocardiography with doppler was performed at each consultation. The mean shortening fraction was of 37% at the BD,the right coronary artery was viewed in the anatomic position in 96% of cases with an anterograde flow.All the patients have had a postoperatory cardiac CT scan. At the control scan,3 patients had a reimplanted right coronary artery appearing of a small caliber ,one of them measured had a 2.5 mm diameter. For a patient,the evolution was towards a complete ostial stenosis of the right reimplanted coronary artery. That patient had a pulmonar atresia and received an ARCA reimplantation surgery associated to a pulmonary valve replacement and of a residual IAC closure.In preoperative the right coronary artery was described as small caliber with a diameter not exceeding 2 mm in its inter-aortopulmonary course.

The postoperative history had been simple. The postoperative TTE showed a permeable and well perfused coronary artery,an RV TAPSE at 18 mm,a well-functioning bioprosthesis and an absence of residual shunt.2 years after the surgery,the patient had a syncopal unease associated with effort-related chest pains .The check-up performed (ECG,Holter-ECG,troponins, TTE, stress test) proved to be negative. The cardiac CT scan showed a small caliber but permeable ostium .Unfortunately, the patient then aged 18, had the same clinical presentation 18 months later which lead us to perform a coronary angiography (image 4).That one has revealed a dominant left coronary system,an astial stenosis of the reimplanted right coronary with an arterial collaterality of the right coronary system. There has not been any painful relapse since.

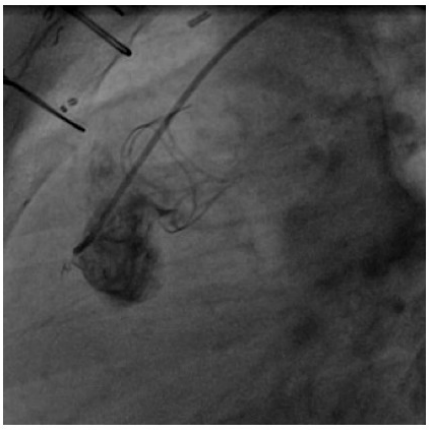

Figure 6. Selective right coronary artery angiogram in left lateral view: complete ostial stenosis in left lateral view, biological pulmonary valve in place.

5 patients have had a myocardial scintigraphy and 3 have had a postoperative coronary angiography.1 scintigraphy showed sequels of a myocardial ischemia but those existed postoperatively.75% of the patients have had a postoperative stress test.3 were sub-maximal. There was no re-operation. However,a patient had an automatic implantable defibrillator placed for recurrent syncopes in the context of an arrhythmogenic right venticular dysplasia.

2.8 Mortality :

There was one death 3 months postoperatively. It is the youngest , operated on at 2 months old, at 3,6 Kg. That child was followed up since antenatal period for fetal

arrhythmia an the ARCA diagnosis was made at birth by echocardiography. No other lesion was associated besides a persistant oval foramen. It was then decided to perform an anti arrhythmic medical treatment,
the child however died suddenly at home and underwent in emergency a direct reimplantation surgery with closure of the IAC (Figure 7)

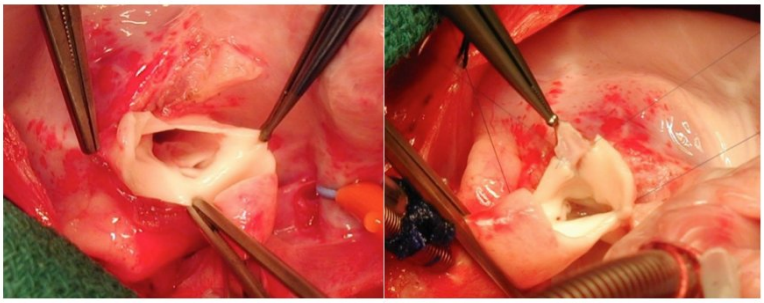

Figure 7. Intraoperative images showing the ostium of the ARCA in the posterior left sinus and its reimplantation in pallet in the anatomic position after transverse aortotomy by double-armed continuous sutures .

3.Discussion :

The exact physiopathological mechanisms of clinical signs and sudden deaths of patients with ARCA are not well known(6).Several hypothesis have been developed in the literature in order to explain the myocardial ischemia and the cardiac rhythm disorder caused by ARCA with ADS (7).The symptoms appear most of the time at the pea of an effort or following immediately and are likely to be the consequence of an external compression of the right coronary artery in its inter-aortopulmonary portion (8). However,even during a prolonged and intense effort, the pulmonary artery blood pressure remains inferior to the one in the coronary artery. Moreover,in a 2003 study, Angelini and al have demonstrated that the course remains "compressed" even in the phases of the cardiac cycle when the pulmonary artery trunk is distant (9). It seems that the intramural course is the real responsible of the symptomatology (22). Several studies have shown that the intramural portion has a smaller intraluminal circumference. Actually, during the effort, the aorta is subject to a parietal tension increase,and through this mechanism,the intramural course is compressed,particularly during the systole (23).This is especially true regarding the valve commissure (10).In an ARCA,the right coronary artery always connects at a non orthogonal angle to the aortic wall. This angle may be more or less acute and associated

to a "king-king".The ostium itself has most of the time in a"flute beak" shape.It may also be stenotic and even punctiform. Sometimes an obstructing membrane is found.
The aortic valve prosthesis may as well cause a partial occlusion of this ostium, especially when it is para-commissural (10).If the first descriptions are from series of autopsy series(11), more and more studies tend to determine the exact prevalence of the AACCA and therefore of the ARCA, from series either of coronary angiograms or x-ray images. The ARCA seem therefore to affect 0,06 to 0,9% of the population.In 2013,Angelini published a MRI screening study on a population of young adults in which he found the same 0,6% of ARCA (12).The ARCA are 6 to 10 times more frequent than the ALCA but are most often asymptomatic.and the ALCA are responsible of nearly 85% of sudden deaths in AACCA (13). In their case series,Maron and al estimated at 0,61 in 10000 persons per year the risk of sudden death in high level athletes,with 17% attributed to AACCA(3) .Brothers and al,through a similar analysis,with a prevalence of AACCA of 0,6% in the general population,and considering the ARCA 6 times more frequent than the ALCA, came to the conclusion that the risk of sudden death in patients who had an ARCA,was of 0,004% (7).This risk has been described as 80 times higher in athletes than in the general population and the athletes afflicted by an AACCA have been excluded from competitions. Finally,the correlation between coronary anomaly and coronaropathy is uncertain but the presence of a coronaropathy could be an aggravating factor of the AACCA.Conversely,an AACCA would increase the risk of coronaropathy (14).
However no predictive risk factor for sudden death could be highlighted and it appears impossible to determine the risk of sudden death in the general population.
The only accepted risk factor is the age inferior to 30 years old at the time of the diagnosis.
The symptomatology of the ARCA is often non-specific and should be further developed. Many patients are diagnosed during a recovery from sudden death.
Most of them declare themselves asymptomatic then before the episode which most often happens during the effort. Some patients are diagnosed incidentally although the motivation behind the tests performance having lead to the diagnosis does question a real asymptomatology.Generally,non-specific chest pains,palpitations,faintness or syncopes are described (1).The electrocardiogram and the holter-ECG are generally normal or within normal ranges.It is possible however to find large T-waves in the V1,V2 and V3 leads .
For some,the cardiac rhythm disorders enable the diagnosis,(ventricular fibrillation, ventricular tachychardia).Similarly, stress tests are electrically and clinically negative ,in the literature,less than 10% of operated patients had a positive stress test (7).
The diagnosis is essentially based on the two-dimensional echocardiogram with colour

doppler. It highlights the anomalous connection,the intramural and inter-aortopulmonary course. The echocardiogram also enables to quantify the ventricular function. In order to confirm the diagnosis,the heart CT scan is the preferred examination,it enables to determine the length of the inter-aortopulmonary course,the intramural course and the ostial anatomy. The latest generations allow to reduce to the maximum the irradiation particularly in the pediatric population (15).

The MRI offers the advantage of assessing the viability and the myocardial ischemia and to eliminate the differential diagnosis of myocarditis (4).

Some authors suggest that it would enable to highlight a myocardial fibrosis sign of a recurrent chronic ischemia and which might be the predictive sign of ventricular rhythm disorders. (16).The coronary angiography is almost not longer used to perform a diagnosis. The myocardial scintigraphy having for purpose to unmask the ischemia lesions is often little contributive.

Facing the physiopathological and diagnostic uncertainties that the AACCA generate,the therapeutic management is very much a topic of debate. If the ALCA seem to obtain a consensus regarding their surgical management,the same does not apply to the ARCA. Moreover,the age at the time of the diagnosis of an ARCA seems for some teams a decisional factor for choice or not of the surgical management (17). The drug therapies by Beta-Blockers or inhibitors of the conversion enzyme have been reported as predictive of the risk of sudden death. Besides,most of the teams contraindicate the practice of sport.However,the restriction of physical activity seems difficult to obtain in the pediatric population and may have physical consequences (overweight) and psychological ones on the patient and his family (18). At this time, anon-operated patient is followed up in our center. It is a 14 years old girl carrier of the Ebstein disease. The ARCA diagnosis has been made when he was 5 years old in TTE and confirmed by a heart CT scan. She never had chest pains nor cardiac rhythm disorders. At the EGC,there is an incomplete right bundle branch block. The cardiac stress test was negative. The echocardiography shows a right ventricular dysfunction linked to the Ebstein disease. A contraindication to sport was implemented. In our center,we contraindicate the practice of competitive sport for all patients who have an AACCA non-operated,the patients and their family are informed of the signs requiring an emergency consultation and a medical follow-up.

Some case series of ARCA treated by interventional catherism may be found in the literature. It is most often ARCA without IAC with ostial stenosis. Different surgical techniques have been suggested in the surgery management of the ARCA with IAC.

In order to protect for the external compression by the pulmonary artery trunk, some

have suggested to proceed to a lateral or anterior translocation of the pulmonary artery trunk. Some authors associate an ostioplasty to that (19,20). For the adult patients, the coronary bypass surgery may be offered with a proximal ligation in order to suppress a competitive flow. When a coronaropathy is associated, this may be more justified but this technique on young adult without coronaropathy is controversial because the distal network is then entirely dependant on the quality and the sustainability of the bypass.The use of the thoracic artery is preferred as it preserves the potential of growth and would better resist to the development of artherosclerosis. However, the implementation of a bypass in children poses various problems of which the one of the revision surgery. The "unroofing"technique is the most widespread,it has been described by Mustafa and al in 1981 and consists in excising the wall of the intramural segment. Difficulty arises when the intramural segment is long and retro-commissural,some authors proceed to a detachment of the commissure but this may lead to long-term aortic insufficiency. This is why a neo-ostium creation could be suggested. Unfortunately, even though very promising, this technique has shown cases of long-term aortic insufficiency and of residual ischemia.The direct reimplantation in the anatomic position is the only technique enabling to overcome all issues related to ostial anomalies,inter-aortopulmonary course end intramural course. It consists in cutting the coronary artery at its point of anastomosis to the aortic wall and to reimplant it in the anatomic position. Quite often,an ostioplasty is associated in order to enlarge this one either by a posterior incision or by a patch enlargement technique. In our center,we have developed a direct reimplantation technique without transverse aortotomy with very good postoperative results. The main difficulty that we find before the aortic clamping is the implementation of the cardioplegia. No patient operated with this technique presented any secondary aortic insufficiency nor underwent any reintervention. It is difficult to establish the long term result of surgical procedures,the literature reporting only case series with few patients, operated using different surgical techniques and with sometimes an absence of distinction between the ALCA and the ARCA. However,no immediate death has been reported. The postoperative complications described are those of any cardiac surgery (postoperative bleeding, pericardial effusion, pneumopathy...) and are generally inferior to 20%.However,these surgeries are most often performed on young patients without comorbidity.It is likewise in our whole serie. The only death that occurred was of a toddler who had suffered from heart rhythm abnormalities as a fetus, had presented a preoperative biventricular dysfunction (FR 21%) after a recovery from sudden death. The described reoperations concern the patients presenting an aortic defficiency. most often in the afternath of the surgical technique by unroofing with

reimplantation of the commissure (20). No patient of our serie was reoperated. For the patient presenting an ostial stenosis a repermeabilisation technique is debated because the stenosis is relatively short and the downstream network permeable, belonging to the left coronary artery. The myocardial viability assessed in that direction shows an absence of ischemic sign. It must be reminded that this patient has a dominant left network.It may be noted that the surgical treatment has replaced over the past
few years the medical treatment and that the treatment strategy is not the same as the one in centers that have a solid experience in congenital cardiac surgery.Over the years, we have observed the progress of the number of patients sent to surgery .All the patients seen in consultation have accepted the surgical management,most of them expressing clearly their fears and even an anxiety depression syndrome about the risk of
sudden death. It is easier to perform a preoperative coronary angiography in adults. The most often chosen technique is the bypass and a procedure is often associated. The hospitalization period is often longer. The only widely admitted risk factor of sudden deaths of patients with an AACCA is being under age 30. In our study,patients were older than 30 at the time of the intervention, which represented one third of the population. They were all symptomatic with a risk of sudden cardiac death. 6 patients had had a non significant preoperative coronary angiography with a non significant coronaropathy. The period before intervention was of 9,6 months compared to 22,9 months for the patients under age 30. There has been 1 lost to follow up. The average follow-up time was 26,9 months. At the LFU , 2 patients were NYHA II . 5 had returned to a physical activity and 3 were complaining of recurrent chest pains. 8 patients underwent a stress test at the DDN
one of which one submaximal for a decrease of the ST segment.

Table 2. Comparison between patients operated before and after age 30

	< age 30 (n = 20)	**>age 30 (n = 10)**	**p-value**
H/F	18/2 = 9	6/4 = 1,5	
Mean weight (kg)	48,6	77,8	
Sport activity	12	2	
Sudden death	4	1	
Stress test	8	6	
Period before intervention(months)	22,9	9,6	p = 0,01
ECC duration	52,4	51,7	p = 0,46
Clamping duration	30,1	28,7	p = 0,40
Associated gesture	4 (3 CIA, 1 PVR)	1 (AVR)	
Hospitalization period(days)	7,5	7,6	p = 0,58
Postoperative complications	3	2	
< 30 days			
Follow-up (months)	24,9	26,9	p = 0,45
NYHA I at the LFU	17 (n = 19)	7 (n = 9)	
Beta-blockers at the LFU	11 (n = 19)	6 (n = 9)	

Comparing the 2 populations (Table 2)
you can see that the number of sudden deaths is higher in the under age 30 , 1 patient under age 10 presented a recovered sudden death like the patients over age 30, we may thus consider that the age group is between age 10 and age 30.The period between the diagnosis and the intervention is shorter in those over 30. Indeed, we have had a more wait-and-see attitude with the youngest with implementation of a physical activity restriction and a contraindication to sport associated with a regular follow-up until the completion of paraclinical examinations (heart CT scan, stress test) in the right conditions. There is no significant difference between these two groups and the surgical outcomes are superposable, in the over age 30 the disappearance of anxiety regarding the diagnosis which seems to improve the quality of life of patients and the patients find themselves in better health but this is probably related to the regular medical follow-up.
However, some of them complain about chest pains but those are described as different from the preoperative pains (parietal pains) . 1 patient only was hospitalized several times for angor but this patient is diabetic,hypertensive and overweight. he presented

preoperatively a coronaropathy with non significant stenosis of the right side.This one has evolved towards a right and left coronaropathy without other necessity than a medical treatment. The remote follow-up implies a minimum annual consultation, with completion of a heart CT scan and a holter-ECG immediately postoperative then followed by a regular echocardiography. The return to physical activity and to competitive sport is authorized 6 months after surgery. Several teams have suggested the use of a decision algorithm for the AACCA management,the most complete is the one used by the team of the Texas Children Hospital (19). The ARCA with IAC must be treated surgically when there are symptoms or ischemic signs at the paraclinical examination, when there is an intramural course or ostial anomalies. The diagnosis has a potential psychological impact on the patient's quality of life and on his family.

4.Conclusion:

The surgical management of the ARCA with IAC remains controversial especially in adults over the age of 30,our study shows a good surgical result of the aortic reimplantation and describes the results of the reimplantation technique without transverse aortotomy. We think that only direct reimplantation enables to restore permanently the anatomy of the coronary artery and authorize the return to physical activity even in competition. The ARCA with IAC are always associated with an intramural course and ostial abnormalities.
In that sense, besides rare cases of incidental findings in the subject over age 50 who may accept the low but non-null risk of sudden death and restrict his physical activities, any patient diagnosed in our center is sent to the surgeon.
This study is a retrospective unicenter study of 30 patients operated by direct reimplantation these past 15 years of whom the results confirm the necessity of the surgical management of these patients, the long-term evolution must however be assessed, in particularly as regards of the evolution or the appearance of a coronaropathy on this reimplanted right coronary. Indeed,any operated coronary ,whatever the technical cause or the cause presents a risk of evolution in itself and must be monitored.

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