ALCAPA – a need for guidelines for managing the adult type

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Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) or Bland-White-Garland syndrome is a rare congenital coronary malformation, accounting for 1 of 300.000 live births and 90% mortality if untreated in infancy[1]. However, in recent years, with the availability of newer, better imaging methods, an increasing number of cases in an older cohort have been identified[2].

Case Report

A 15-year old boy was referred to a University Hospital, for investigation of a recently en passant discovered heart murmur. He felt completely healthy, was a member of the local ice-hockey team and was playing at least 5 times a week without any symptoms. Physical findings consisted of a physically fit young man with normal breathing, normal heart rate and normal blood pressure. The only abnormal finding was a harsh systo-diastolic murmur grade III/IV, localized at the left sternal boarder and in the apex region, but not heard on the back.

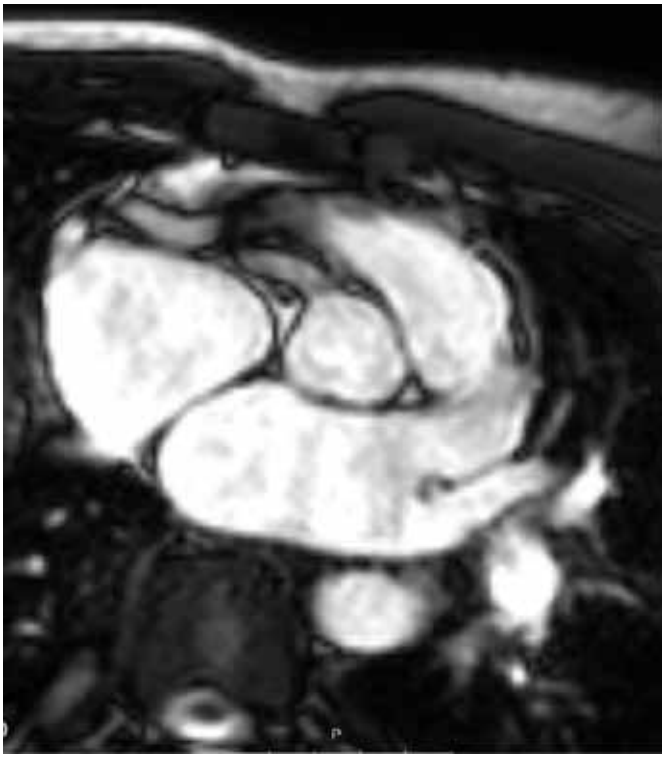

Figure 2: Short axis steady-state free-pressesion image at the level of the origin of the right coronary artery. The right coronary artery originates at it usual location whereas the left coronary artery is not identified in this image.

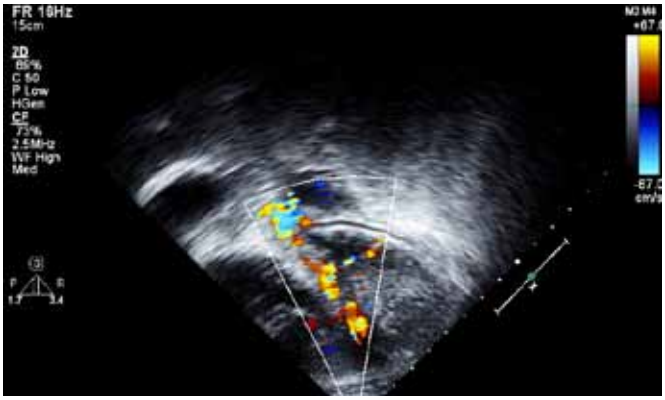

Figure 1a: Apical 4-chamber view showing multiple, predominantly diastolic, abnormal septal color flows indicating septal perforator coronary arteries.

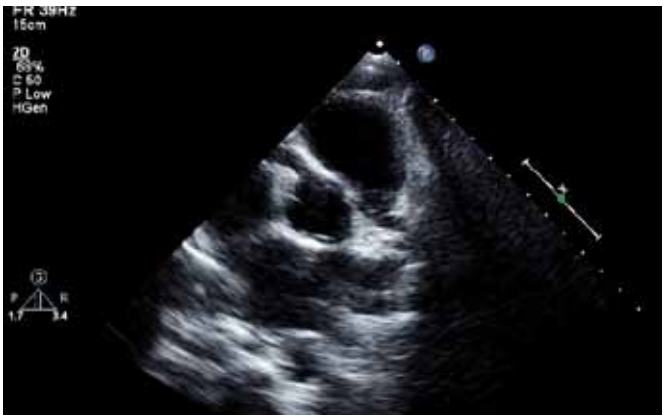

Figure 1b: From parasternal short-axis view, a markedly dilated right coronary artery (RCA) can be seen.

The electrocardiogram showed normal sinus rhythm, no signs of (ventricular) hypertrophy, myocardial ischemia or other pathological findings. The first transthoracic echocardiographic examination showed a slightly enlarged left atrium, trivial mitral valve regurgitation and a normal sized left ventricle with normal ejection fraction. Within the interventricular septum, multiple predominantly diastolic abnormal color flows were detected (Fig 1a). From parasternal short-axis view, a markedly dilated right coronary artery (RCA) could be shown but the origin of the left coronary artery (LCA) was not identified (Fig 1b). An abnormal retrograde blood flow from the pulmonary artery with low diastolic velocity was though detected and these findings led to the suspicion of plausible congenital coronary artery malformation.

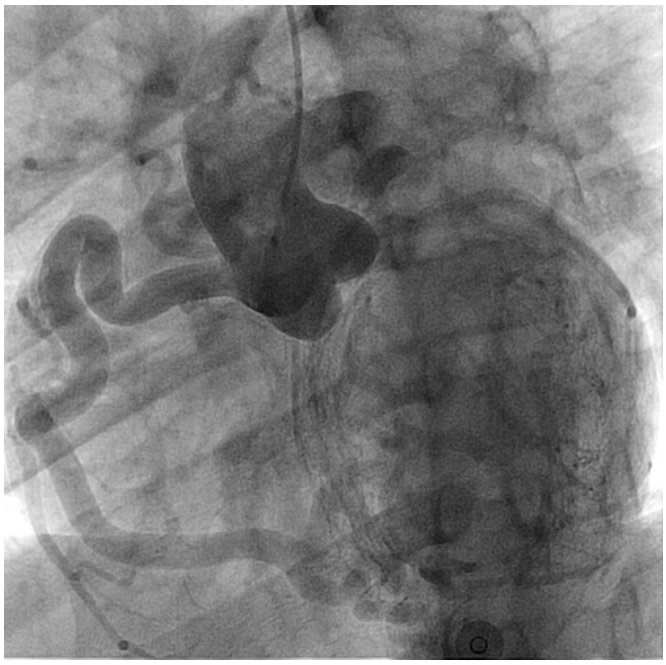

Figure 3: Coronary angiography confirmed a dilated, tortuous RCA with extensive collaterals filling the left coronary artery (LCA). The left main stem originated from the main pulmonary artery – i.e. an anomalous origin of the LCA from the pulmonary artery (ALCAPA) with retrograde flow into the main pulmonary artery.

Further evaluation with treadmill test showed good exercise capacity without any evidence of myocardial ischemia. Myocardial scintigraphy during exercise showed a minimal reduced distribution of radioactivity in a small area in the anterior wall of in the left ventricle which increased slightly during exercise. MRI identified a wide, tortuous RCA, a normal left ventricle with homogenous myocardium and an ejection fraction of 65% without any regional reduced motility (Fig 2). Evaluation with late gadolinium enhancement was not performed. The MRI study was inconclusive regarding visualizing the origin of the LCA origin and subsequent coronary angiography was implemented.

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Discussion

Pathophysiology

Anomalous origin of the left coronary artery from the pulmonary artery is well tolerated in fetal life as the pressure in the pulmonary artery is equal to aortic pressure which results in antegrade flow in LCA. During the first two postnatal months, the pressure in the pulmonary artery falls, leading to decreasing flow in LCA. Finally the flow in LCA reverses which consequently results in myocardial ischemia. This represents a very critical period and 90% of untreated cases die within the first months or in the first year of life[3].

However, some patients develop collaterals between the RCA and LCA during the period when pulmonary arterial pressure gradually decreases. If well-established collaterals are formed, the patient will develop the adult form of ALCAPA[1]. These patients are initially asymptomatic but in the long run, the collateral circulation from RCA to LCA becomes not sufficient and leads to chronic left ventricular subendocardial ischemia, which in turn may result in malignant arrhythmias and left ventricular dysfunction. It is known that up to 80-90 % of the patients in this group may die from sudden cardiac death[4, 5].

Investigations

The ALCAPA diagnosis used to be more often set post-mortem by necropsy. Nowadays, newer diagnostic methods such as CT and MRI have resulted in more frequent diagnosis of the adult form of ALCAPA. Yau et al in a comprehensive review of 151 adult ALCAPA cases2 found that in the group with average age of 41 years, 14% were asymptomatic. Our present patient did not have any symptoms, not even during heavy physical exercise, but he can be considered more as an adolescent than an adult. In the review study from 2011 the authors found that 21 ALCAPA patients had a total of 51 uncomplicated pregnancies, a condition that requires good physical ability.

The only clinical sign in our patient was a harsh systo-diastolic, but not continuous murmur. In the study of Yau et al 87% of the patients had murmurs, 71% systolic. The murmurs were predominantly localized to the left sternal border or apical region, as in our patient. Seventeen percent of the 151 ALCAPA cases had ventricular arrhythmias, syncope or sudden death as their first presentation. At the time of presentation, 66% reported angina, dyspnea, palpitations or fatigue – in patients with cardiac disease rather common and vague symptoms[2]. Echocardiographic diagnostic criteria for ALCAPA have been established including dilated RCA, with flow from LCA to the pulmonary artery and signs of collaterals in the interventricular septum6. An important fact is though that echocardiography is not a reliable method for ALCAPA diagnosis. In the study of Yau et al 76 patients had echocardiography and only 46% were diagnostic/suggestive for ALCAPA, 44% were abnormal but did not indicate ALCAPA and 10% were normal[12].

However, in recent years, other non-invasive imaging modalities such as multidetector computed tomographic (MDCT) angiography and magnetic resonance imaging (MRI), have emerged. Both methods usually enable visualization of the left coronary artery arising from the main pulmonary artery. MRI may also further show the reversed flow from LCA into the pulmonary artery and assess the myocardial viability by gadolinium, however not performed in our case with normal left ventricular performance. A recent review has demonstrated an increasing number of non-invasive ALCAPA diagnosis since year 2000, which correlates with an increasing incidence of ALCAPA in the adult population[2].

Treatment

There are still controversies about when to correct adult ALCAPA patients but the vast majority recommends surgical treatment in asymptomatic adults if only moderate chronic ischemia is present[1, 2, 7, 8, 9]. But still, especially in asymptomatic older patients, the risk-benefit ratio of surgery is unclear[9].

There are several different surgical procedures for ALCAPA including direct reimplantation of the LCA into the aorta, ligation of the anomalous LCA, various forms of bypass grafts from the aorta, subclavian-LCA anastomosis, and Takeuchi procedure (creation of an aortopulmonary window and an intrapulmonary

tunnel that baffles the aorta to the ostium of the anomalous LCA)[10].

It is obviously desirable to reimplant the LCA into the aorta to achieve a two-coronary circulation system, but this is not always possible to achieve if the distance between the origin of the anomalous LCA and the aorta is too long. In our 15-year old patient, the distance was too long for reimplantation resulting in controversial opinion about how to surgically treat or NOT treat this patient.

This case raises many questions as how to manage the asymptomatic adolescent/adult patient with ALCAPA:

The principal investigation
- Is MRI the gold standard for investigation of ALCAPA- should always gadolinium injection be included in the examination?
- Should both CT and MRI be performed?
- Myocardial scintigraphy, echocardiography or CMR perfusion during stress - should these modalities be included in the investigation of a young adult patients?

Counseling on physical activity
- If the patient is completely asymptomatic and show no signs of myocardial ischemia on treadmill test, should he/she be restricted from physical activity?
- If not surgically treated – what about life style recommendations?

Indications for surgical treatment
- What are the indications in the young for surgical interventions in patients with minimal myocardial disturbances?
- When should the intervention be initiated?

Conclusion

Parallel with the development of improved imaging methods, an increasing number of asymptomatic patients with ALCAPA in all ages will be identified - guidelines are needed to assist optimum management of this “new” cohort of patients.

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References

1. Peña E, Nguyen ET, Merchant N, Dennie C. ALCAPA Syndrome: Not Just a Pediatric Disease. RadioGraphics 2009; 29: 553-565.
2. Yua J, Singh R, Halpern EJ, Fischman D. Anomalous Origin of the Left Coronary Artery in Adults: A Comprehensive Review of 151 Adult Cases and A New Diagnosis in a 53-Year-Old Woman. Clin Cardiol 2011; 34(4): 204-210.
3. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. Circulation 1968; 38: 403–425.
4. Alexi-Meskishvili V, Berger F, Weng Y, Lange PE, Hetzer R. Anomalous origin of the left coronary artery from the pulmonary artery in adults. J Card Surg 1995; 10: 309–315.
5. Berdjis F, Takahashi M, Wells WJ, Stiles QR, Lindesmith GG. Anomalous left coronary artery from the pulmonary artery: significance of intercoronary collaterals. J Thorac Cardiovasc Surg 1994; 108: 17–20.
6. Yang YL, Nanda NC, Wang XF, Xie MX, Lu Q, He L, Lu XF. Echocardiographic diagnosis of anomalous origin of the left coronary artery from the pulmonary artery. Echocardiography 2007; 24: 405–411.
7. Tian Z, Fang LG, Liu YT, Zhang SY. Anomalous origin of the left coronary artery from the pulmonary artery detected by echocardiography in an asymptomatic adult. Intern Med 2013; 52: 233-236.
8. Komócsi A, Simor T, Tóth L, Szabados S, Mágel F, Pintér T, Aradi D, Kónyi A, Horváth IG, Papp L. Magnetic resonance studies in management of adult cases with Bland-White-Garland syndrome. Int J Cardiol 2007; 123: e8–e11.
9. Kottayil BP, Jayakumar, K, Dharan BS, Pillai VV, Ajitkumar V, Menon S, Sanjay G. Anomalous Origin of Left Coronary Artery From Pulmonary Artery in Older Children and Adults: Direct Aortic Implantation. Ann Thorac Surg 2011; 91: 549 –554.
10. Kandzari DE, Harrison JK, Behar VS. An anomalous left coronary artery originating from the pulmonary artery in a 72-year-old woman: diagnosis by color flow myocardial blush and coronary arteriography. J Invasive Cardiol 2002; 14: 96–99.
11. Lange R, Vogt M, Horer J, Cleuziou J, Menzel A, Holper K, Hess J, Schreiber C. Long-term results of repair of anomalous origin of the left coronary artery from the pulmonary artery. Ann Thorac Surg 2007; 83: 1463–1471.