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CASE REPORT

T. Schneider
H. Rickli
V. Gliech
M. Maeder

# Bland-White-Garland syndrome and atrial septal defect –

## Rare Association and diagnostic challenge

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Dr. med. Micha Maeder (✉)
Division of Cardiology
University Hospital Basel
Petersgraben 4
4031 Basel, Switzerland

Dr. med. Tino Schneider
Dr. med. Hans Rickli
Kantonsspital St. Gallen
Department Innere Medizin
Fachbereich Kardiologie
Rorschacherstr. 95
9007 St. Gallen, Switzerland

Dr. med. Volker Gliech
Humaine Klinikum
Bad Saarow/Fürstenwalde
Pieskowerstr. 33
15526 Bad Saarow, Gemany

**Summary** We report on a 40-year-old woman referred for evaluation of a cardiac murmur and dyspnea on exertion. The electrocardiogram (ECG) showed incomplete right bundle branch block, and echocardiography revealed a large atrial septal defect (ASD, ostium secundum type) with dilated right-sided heart chambers. At cardiac catheterization, a large left-to-right shunt (78% of the pulmonary blood flow) was found, and surprisingly, the additional diagnosis of anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) was established. After ASD closure and left coronary artery ligation with implantation of a vein graft to the left anterior descending artery, she had an uneventful 18-years follow-up.

We discuss the interaction of the two associated conditions, and based on the herein reported unusual combination, we highlight typical features of non-invasive examinations including auscultation, ECG, and echocardiography in adult patients with ALCAPA.

**Key words** Shunt – atrial septal defect – coronary anomaly – non-invasive diagnostics

## Inroduction

Anomalous origin of the *L*eft *C*oronary *A*rtery from *P*ulmonary *A*rtery (ALCAPA or Bland-White-Garland syndrome) is a rare congenital coronary artery anomaly occurring in approximately one in 300 000 live births [3, 22]. Ischemia of the myocardial territory supplied by the left coronary artery occurs as flood flow in the left coronary artery is reversed after birth. Perfusion of the left ventricular myocardium therefore depends on collateral flow from the right coronary artery. Up to 90% of children with ALCAPA die during the first year of life because of myocardial ischemia and left ventricular failure. Survival until adulthood is rare and depends on pre-existing or rapidly developing collateral vessels between the right and the left coronary artery [19, 22, 28].

Whereas ALCAPA usually occurs as an isolated lesion, there are few reports on its association with other intracardiac defects [20, 24]. We herein describe the case of an adult patient undergoing cardiac catheterization for evaluation of large atrial septal defect (ASD), in whom the diagnosis of co-existing ALCAPA was established. Our report focuses on the clinical impact, the diagnostic challenges, and the therapeutic implications encountered in the setting of this unusual combination.

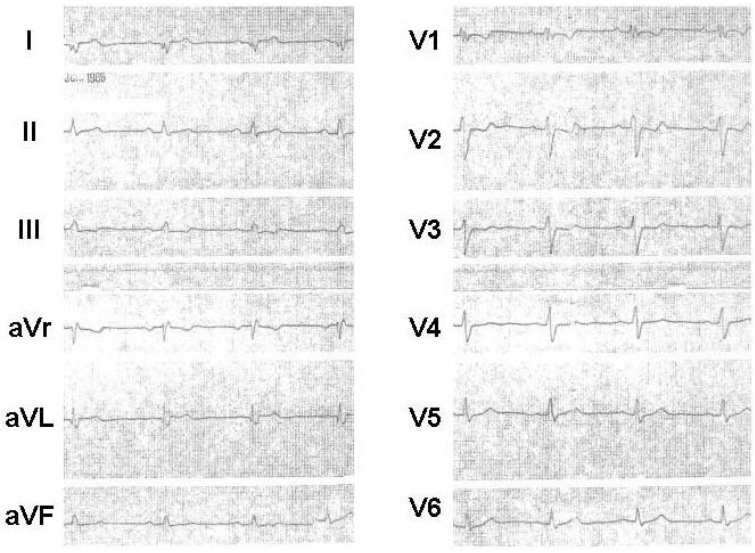


**Fig. 1** Electrocardiogram (paper speed 50 mm/s) showing sinus rhythm with incomplete right bundle branch block and negative T-waves in leads V1–3. There are small Q waves in leads I and aVL, and in lead II T waves are negative

## Case report

In 1986, a 40-year-old woman was referred because of progressive dyspnea and a cardiac murmur, which had been known for years. Otherwise, the patient's medical history had been unremarkable. After an uneventful pregnancy she had given birth to a healthy boy 15 years ago. At presentation the patient was in New York Heart Association (NYHA) class III. Physical examination revealed fixed splitting of the second heart sound. Both a systolic and a diastolic murmur were audible at the left sternal border. The electrocardiogram (ECG) is shown in Fig. 1. Transthoracic echocardiography revealed a large ASD (ostium secundum type, diameter 14 mm) and dilated right-sided heart chambers. As operative ASD closure was planned, the patient underwent cardiac catheterization. Mean pulmonary artery pressure was 27 mmHg, and a large left-to-right-shunt was calculated by dye dilution (78% of the pulmonary blood flow). Mild anterolateral hypokinesis of the left ventricle was noted, the global left ventricular ejection fraction being normal. At coronary angiography, a left coronary ostium could not be identified, but injection into the right coronary artery revealed a very large vessel with retrograde collateral filling of the left coronary and entry of the contrast agent into the pulmonary artery (Fig. 2). Thus, the surprising diagnosis of ASD combined with ALCAPA had been established. The patient underwent ASD patch closure, left coronary artery ligation at its origin from the pulmonary artery, and implantation of a vein graft to the left anterior descending artery. She made an uneventful recovery after surgery.

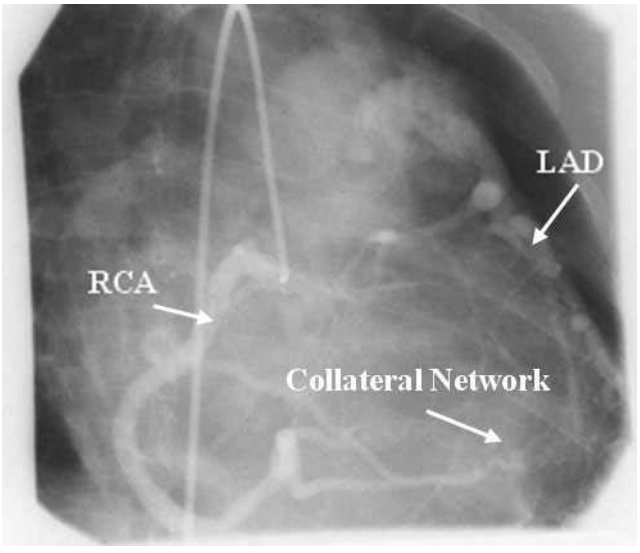


**Fig. 2** Coronary angiography showing a large right coronary artery (RCA) with collateral filling of the left anterior descending coronary artery (LAD) and contrast entry into the pulmonary trunk by a collateral network

Eight-teen years later, at the age of 58 years the patient was in NYHA class II. Exercise capacity was markedly limited by a concomitant neuromuscular disorder. However, repeated echocardiographic follow-up examinations had shown complete normalization of systolic left ventricular function, and the left ventricular end-diastolic diameter had decreased from 43 to 38 mm. Dilatation of the right-sided chambers had resolved, and pulmonary hypertension was absent.

## Discussion

The present example of ALCAPA is unusual with respect to both the patient's less symptomatic course until adulthood and the co-existing ASD with associated diagnostic and therapeutic pitfalls.

Most patients with ALCAPA become symptomatic a few weeks to months after birth, when the physiological neonatal pulmonary hypertension has resolved, and closure of the ductus arteriosus has occurred [28, 29]. Flow in the left coronary artery is then directed towards the low-pressure pulmonary artery leading to a steal phenomenon in the myocardial territory supplied by the left coronary artery [2]. Pre-existing or rapidly developing collateral vessels between the right and the left coronary artery may prevent a fatal clinical course early in life, but the left ventricle remains chronically hypoperfused resulting in a variable degree of left ventricular dysfunction [25]. Nonetheless, long-term survival in patients with ALCAPA without surgical correction has been well documented [1, 4, 7, 9, 10, 14–18, 21, 23, 27].

In our patient, the large left-to-right shunt across the ASD with subsequently elevated pressure and oxygen saturation in the pulmonary trunk may have improved blood and oxygen supply to the left coronary artery by attenuating retrograde left coronary flow, and hence the beneficial interaction of two potentially hazardous conditions may have contributed to the long asymptomatic period including an uneventful pregnancy. Regarding the significant left-to-right shunt with right ventricular overload, the indication for ASD closure was evident [5]. However, operation without knowledge of the coronary anatomy might have been fatal, as the sudden occurrence of severe left ventricular ischemia due to an abrupt reduction of pulmonary artery flow and oxygen saturation following ASD closure with subsequent coronary steal from the left coronary artery would have been a very probable scenario. An interesting observation underscoring our speculation comes from the report on a 4-month-old infant with undiagnosed ALCAPA undergoing repair of ventricular septal defect (VSD), in whom severe left ventricular failure occurred after VSD closure. After echocardiographic detection of ALCAPA and reimplantation of the left coronary artery into the aorta, the patient could be weaned from cardiopulmonary bypass and recovered [24]. In contrast to ASD, patients with VSD often have marked pulmonary hypertension, which may even become fixed, and therefore a VSD has certainly a more significant impact on co-existing ALCAPA as compared to an ASD, where volume rather than pressure overload of the pulmonary circulation is present [5]. Nevertheless, in our patient mean pulmonary pressure was somewhat elevated, and hemodynamics may have been perfectly adjusted to and depended on the co-existence of ALCAPA and ASD over years, and thus preoperative coronary angiography probably prevented an adverse outcome. This is a very critical issue, as ASD closure can now be performed percutaneously [26], and coronary angiography is not carried out in all cases.

There are many reports of young adults with yet undiagnosed ALCAPA, who were successfully resuscitated after a cardiac arrest due to ventricular fibrillation, and in whom only a thorough evaluation led to the correct diagnosis [4, 15, 17]. Therefore, we want to stress the importance of typical findings from non-invasive tests in patients with ALCAPA. First, the patient had a systolic murmur as very often heard in patients with ASD [5]. In addition, there was a diastolic murmur, which was attributed to pulmonary regurgitation as the diagnosis of large ASD was evident. However, the murmur has been more likely due to collateral flow from the right to the left coronary artery, which peaks during diastole. Other auscultatory findings in patients with ALCAPA include systolic murmurs of various degrees due to mitral valve regurgitation following ischemia or infarction of the anterolateral papillary muscle or secondary to ventricular dilatation [6].

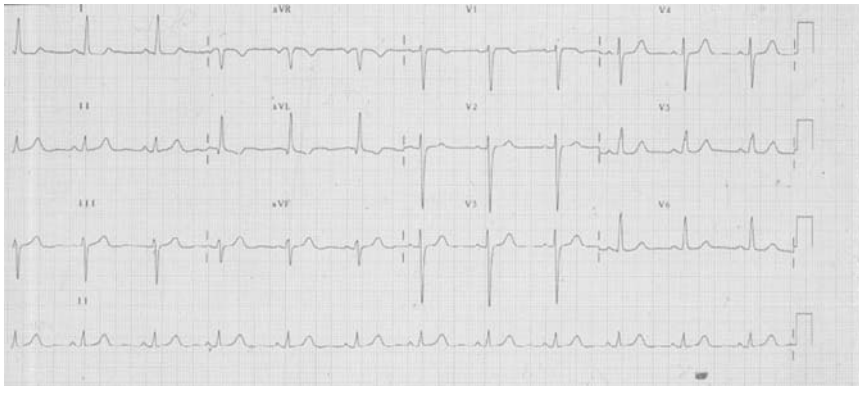

**Fig. 3** Electrocardiogram from a 39-year-old woman with anomalous origin of the left coronary artery from pulmonary artery (case described in reference [16]) showing Q waves and negatives T waves in leads I and aVL. Note that the Q waves are very small, and that signs of left ventricular hypertrophy are absent

Second, patients with ALCAPA often have a characteristic ECG pattern including deep and broad Q waves and/or negative T waves in leads I and aVL and signs of left ventricular hypertrophy [6, 22]. However, classical Q waves are typically less pronounced or even absent in adults as compared to neonates or children with ALCAPA [6]. In Fig. 3, a typical ECG of an adult patient with ALCAPA is shown. In the present case, there were ASD-associated ECG changes, i.e. incomplete right bundle block with T wave abnormalities in the precordial leads, whereas the presence of ALCAPA was not evident. A similar ECG pattern has been reported previously in a 70-day-old boy suffering from both ALCAPA and ASD [20]. In that patient minimal Q waves and negative T waves in aVL were the only ECG signs of ALCAPA [20].

Third, transthoracic echocardiography is a crucial non-invasive tool as it can definitely establish the diagnosis of ALCAPA by demonstration of a large right coronary artery and a mainly diastolic flow from an abnormal vessel originating from the pulmonary trunk [6, 17]. In Fig. 4, the typical echocardiographic aspect of the huge right coronary artery in an adult with ALCAPA is shown, and in Fig. 5, flow from the left coronary artery into the pulmonary artery is visualized. In some cases, collateral flow through the interventricular septum can be seen in the transthoracic study [11]. However, these typical features can be easily missed, especially if another diagnosis is obvious. Other echocardiographic findings seen in patients with ALCAPA, which are non-specific however, include regional wall motion abnormalities, most often localized in the anterolateral wall. Interestingly, ischemia not always occurs in the myocardial territory supplied by the left coronary artery, but also in the infero-posterior wall, as shown by myocardial scintigraphy [13, 18]. The latter finding has been interpreted as a steal phenomenon in patients with large collateral flow [13]. New imagine techniques such as multislice computed tomography and magnetic resonance imaging can accurately visualize the pathoanatomy of ALCAPA [14], but are not always available.

As long as the diagnosis is established, even oligosymptomatic patients with ALCAPA should be operated due to an increased risk of sudden cardiac death [1, 8, 10]. Whenever possible, a two-coronary artery system will be established, either by direct implantation of the left coronary artery into the ascending aorta, or by ligation of the left coronary artery combined with aortocoronary bypass grafting [1, 8]. Isolated ligation of the left coronary artery at its origin from the pulmonary artery has been successfully performed as an emergency life-saving procedure, but is not recommended for routine application as it has been shown to be associated with residual silent myocardial ischemia and a less favorable outcome than the creation of a two-coronary system [8]. Even in patients with significant mitral regurgitation, mitral valve repair or valve replacement is rarely indicated as mitral regurgitation often resolves after revascularization [8]. However, most data on surgical results come from operations in in-

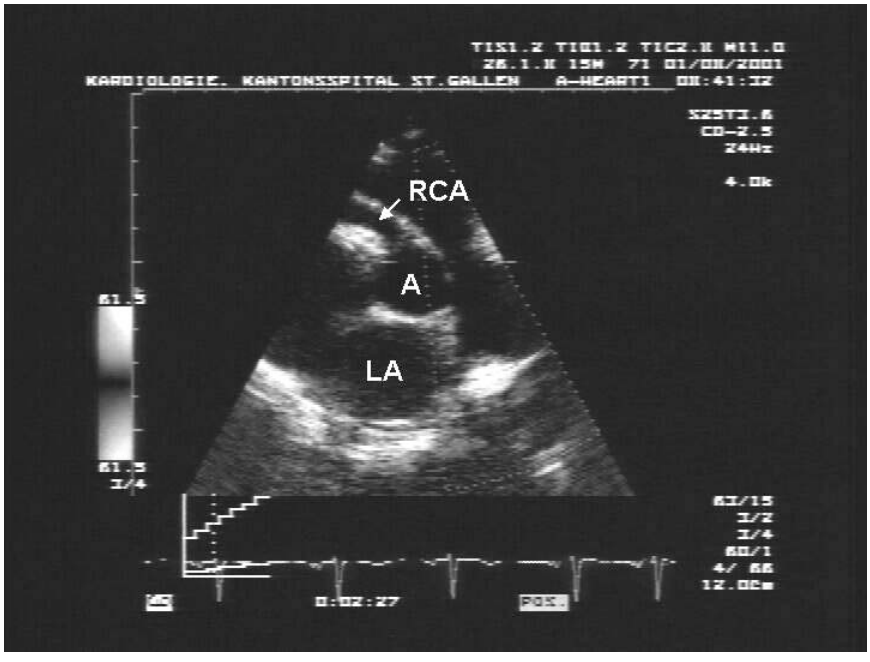


**Fig. 4** Transthoracic echocardiogram (parasternal short axis view) obtained from a 15-year-old male who after a collapse was found in ventricular fibrillation and was successfully resuscitated, showing a huge right coronary artery (RCA) arising from the aorta (A). *PA* = pulmonary artery

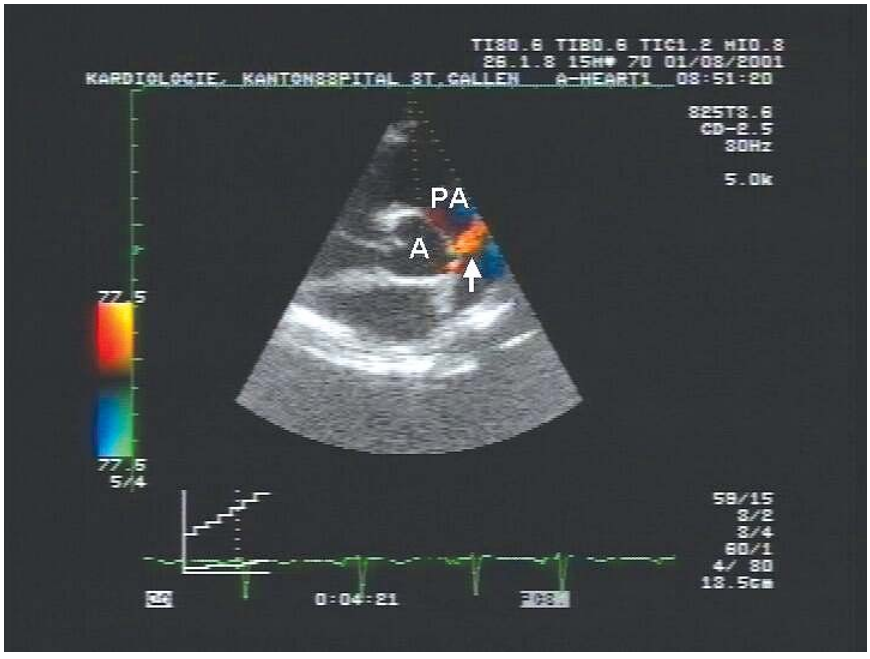


**Fig. 5** Transthoracic echocardiogram (parasternal short axis view) from the same patient showing an abnormal vessel entering the pulmonary trunk (arrow). In this patient, the diagnosis of anomalous origin of the left coronary artery from pulmonary artery was established by echocardiography before transfer to a catheterization center. *A* = aorta; *PA* = pulmonary artery

fants, where complete recovery of left ventricular function, normalization of left ventricular dilatation, and regression of left ventricular hypertrophy within months after reimplantation of the left coronary artery into the aorta has been demonstrated [12]. These changes however, occurred much faster in infants than in older children [12] and may be even slower or incomplete in adults [16]. Therefore the impact of correction of the coronary anatomy on mitral valve regurgitation is hard to predict in adults.

In our patient, combined and simultaneous surgical correction of both ASD and ALCAPA was indispensable as pointed out above. There are few follow-up data in adult ALCAPA patients after correction. The present case is an example for an excellent long-term result after surgery for ALCAPA.

Our case study is limited by the fact that our patient was evaluated almost twenty years ago, and that since then significant progress in echocardiography equipment has occurred. Nevertheless, we feel that the present case is a suitable example to highlight the need for a thorough evaluation of patients with congenital heart disease even if the diagnosis seems to be obvious.

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