The ‘Cauliflower Heart’: a case report of congenital bi-atrial aneurysms causing non-controllable arrhythmia

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Background
Congenital aneurysms of the atrium are very rare malformations. Known complications are therapy-resistant arrhythmias. Different treatments such as medical therapy, electrophysiological ablation, and surgery have been proposed. However, there are no guidelines on treatment.

Case summary
We describe the case of a neonate with bi-atrial aneurysms causing atrial arrhythmia. Arrhythmia was first observed in the 28th week of gestation. Maternal digoxin treatment did not show any effect. After birth, bi-atrial aneurysms were diagnosed and determined as the probable cause of the atrial tachycardia and later of atrial flutter. Antiarrhythmic drug treatment was initiated. However, only frequency control could be achieved. At the age of 7 months, the patient underwent surgical resection. Since surgery, sinus rhythm is present.

Conclusion
Atrial aneurysms are rare malformations, known complications are atrial arrhythmia. If medical treatment fails, surgery correction appears to be indicated going along with low operative risk and a high probability of successful termination of arrhythmia.

Keywords
Atrial aneurysm • Atrial tachycardia • Surgical resection • Case report

Introduction
Atrial aneurysms are a very rare congenital malformation of the heart. The aetiology of atrial aneurysms is unknown1; however, they are often associated with multifocal atrial tachycardia or atrial flutter. No guidelines are available for the treatment of this condition. Some authors state that only aneurysms which cause rhythm problems should be resected,2 others conclude that also asymptomatic patients...
should be treated, as there might be a high risk of thrombus formation and (paradoxical) embolism. In the few existing case reports, there is a consensus that the only sufficient treatment of arrhythmia caused by this malformation is surgery.

Timeline

| Event | Details |
|-------|---------|
| 28th week of gestation | Incidental finding of foetal tachycardia, trial of digoxin treatment without any effect |
| Birth | Multifocal atrial tachycardia, echocardiography shows right atrial mass and atrial septal defect of secundum type, start treatment with propranolol |
| 1 week after birth | Computed tomography angiography: diagnosis of bi-atrial aneurysms |
| 2 months after birth | Atrial flutter, extension of drug treatment by adding Flecainid, only frequency control, no change of flutter |
| 7 months after birth | Surgical resection of the bi-atrial aneurysms, right atrial ablation, atrial septal defect closure |
| 1 year after surgery | Sinus rhythm since surgery |

Case presentation

We present the case of a girl prenatally known for atrial tachycardia which was first diagnosed at 28 weeks of gestation during a routine cardiotocography control. The mother had a past medical history of stomach bypass and two abortions. The actual pregnancy was complicated by an insulin-dependent gestational diabetes. Foetal ultrasound was normal. Maternal treatment with digoxin was started; however, it was stopped 4 weeks later since there was no effect. Pregnancy continued without problems. The girl was born at 37 weeks of gestation. It showed good neonatal adaptation with an Apgar of 8/8/9 and was haemodynamically stable with a heart frequency of 180 b.p.m (It is a score for newborn condition after birth named after Virginia Apgar. Performed at 1, 5, and 10 min after birth. 0 is the lowest, 10 is the maximum. An Apgar of 8/8/9 means good adaption without any problems). Physical examination after birth revealed a normal cardiovascular exam except of tachycardia. Postnatal electrocardiogram (ECG) revealed multifocal atrial tachycardia. Echocardiography showed multiple bi-atrial aneurysms as well as an atrial septal defect (ASD) of secundum type (ASD II). A drug treatment with propranolol was started.

Computed tomography (CT) scan confirmed the diagnosis of four atrial aneurysms (Figure 1), three on the anterior free wall of the right atrium near the tricuspid annulus and one on the anterior free wall of the left atrium next to the pulmonary artery (Figure 1). The patient was discharged from the hospital with frequent follow-ups in our outpatient clinic. At the age of 2 months, the ECG in a routine control revealed an atrial flutter with 2:1 conduction (Figure 2). Flecainid was added to the drug treatment, as this is a well-known drug concept in newborns, resulting in frequency control, however persistence of flutter. An accidental period without Flecainid treatment resulted in a relapse of tachycardia. Due to failure of medical therapy after 7 months, the indication for surgical intervention was given.

Bi-atrial aneurysm resection and ASD closure were performed under cardiopulmonary bypass through a median sternotomy (Figures 3 and 4). The aneurysms were opened and resected at their base. To prevent further atrial flutter and conduction of tachycardia to the ventricle an ablation line was drawn in the right atrium along the junction of the inferior caval vein and the entry of the coronary sinus, to isolate the operation scars from the ventricle. Postoperative course was uneventful with persistence of sinus rhythm. Regular follow-up always showed sinus rhythm without any arrhythmia, the most recent follow-up dates of 1 year after surgery.

A histopathological exam of the resected tissue revealed macroscopically a wedged out wall of the heart to less than 1 mm. Microscopically thin fibrotic and membranous aneurysm walls and trabeculated muscle bridges of atrial myocardium were seen. Additional immunohistochemistry did not show any specificity and no signs of inflammation.

Discussion

Congenital atrial aneurysms are a rare malformation. There is no clear recommendation about best treatment strategy. Diagnosis usually is made by echocardiography. However, angiographic CT scan with 3D reconstruction can facilitate surgical intervention planning. The reconstruction is also helpful to plan resection surgery later on and is possible with low radiation dose.
There are descriptions of complications like pulmonary embolism, thrombus formation, and stroke due to paradoxical embolism in the presence of an ASD. Additionally, the risk of rupture of the aneurysms is not known.

In our case, arrhythmia was the first symptom detected prenatally with only later detection of atrial aneurysms, and medical treatment was initiated rather early.

Following prenatal detection of arrhythmias not responsive to medical therapy subsequent diagnostic revealed multiple bi-atrial aneurysms. A combined antiarrhythmic therapy could just achieve frequency control. Beside the child showed normal growth and development. The reasons for surgical resection were the persistence of arrhythmia beside medical therapy and a potential risk of (paradoxical) embolization because of an additional ASD and therefore risk of stroke. Anticoagulation was evaluated, however, resigned because of bleeding risk. Based on previous reports surgical resection can be considered as a low-risk intervention. In follow-ups the child showed an uneventful postoperative course with normal development and persistence of sinus rhythm.
Lead author biography

Fabienne Stoller, MD, is a paediatrician at the University Children’s Hospital Zurich in Switzerland. She studied medicine at the University of Zurich. She completed her paediatric residency in Switzerland at the hospitals of Yverdon-les-Bains, Biel and the University Hospital of Bern. She was a fellow in paediatric cardiology at the centre for congenital heart disease at University Hospital of Bern. Recently, she rotated to the University Children’s Hospital Zurich to complete her fellowship in paediatric cardiology.

Supplementary material

Supplementary material is available at European Heart Journal - Case Reports online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient’s next-of-kin in line with COPE guidance.

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