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Brief Report

Inverted left atrial appendage in an infant during cardiac surgery

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Abstract Inverted left atrial appendage is an unusual complication associated with congenital cardiac surgery. Inversion of the left atrial appendage may occur during the surgical procedure or afterwards. The left atrial appendage may invert iatrogenically or as a result of the negative pressure during placement or removal of the left atrial vent or during deairing manoeuvres. This event can be life-threatening because of the mass effect of the atrial appendage within the left atrial cavity.

Keywords: Inverted left atrial appendage; congenital cardiac surgery; intraoperative complications; infant

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Inverted left atrial appendage is a rare event during or after congenital cardiac surgery.[1–7] This complication can be life-threatening because of the mass effect of the appendage within the left atrium and because of the associated risk of inflow obstruction of the mitral valve orifice.[6] The inversion of the left atrial appendage is generally diagnosed after cardiac operations; however, the time of occurrence may frequently be during the actual surgical procedure.[1–3] It frequently presents as an asymptomatic mass indistinguishable from thrombus formation or tumour within the left atrial cavity. In this report, we present the intraoperative diagnosis of inverted left atrial appendage in an infant, who underwent repair of ventricular and atrial septal defects. This complication was incidentally diagnosed using intraoperative transoesophageal echocardiography examination after weaning from cardiopulmonary bypass.

Case report

A 7-month-old girl with Down's syndrome and systemic pulmonary hypertension was referred to our clinic for surgical repair of a large perimembranous ventricular septal defect and a secundum atrial septal defect. The intraoperative transoesophageal echocardiogram confirmed the diagnosis of atrial and ventricular septal defects with a normal left atrial cavity. After systemic heparinisation, cardiopulmonary bypass was established at 28° and a left atrial catheter was inserted through the right upper pulmonary vein to decompress the left atrial cavity. Negative suction was applied to this left atrial sump to decompress the left heart throughout cardiopulmonary bypass. Through a right atriotomy incision, the patient underwent combined ventricular septal defect and atrial septal defect closure using pericardial patches. The patient was rewarmed and the left atrial sump was removed immediately after separation from cardiopulmonary bypass. The aortic cross-clamp and cardiopulmonary bypass times were 32 and 72 minutes, respectively.

The haemodynamic status of the patient was stable after weaning from cardiopulmonary bypass. The postoperative transoesophageal echocardiogram showed no residual septal defects; however, it did reveal a 0.9×0.9 cm mobile mass within the left atrial cavity (Fig 1, left panel), which moved towards the mitral valve annulus during diastole (Fig 1, right panel). The lesion was first considered to be an intracardiac thrombus, but this was thought to be unlikely because of the intraoperative systemic heparinisation and

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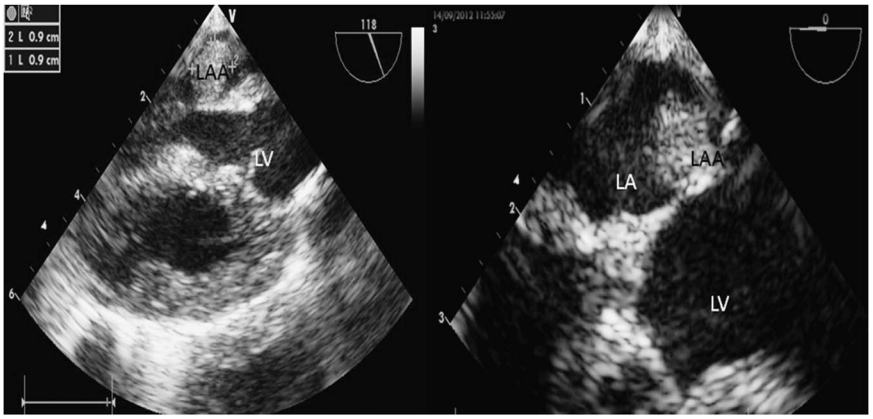

Figure 1.
Intraoperative transoesophageal echocardiographic images show inverted left atrial appendage – left panel. The mass was moving towards the mitral annulus during cardiac cycle – right panel. LA = left atrium; LAA = left atrial appendage; LV = left ventricle.

elevated activated clotting time throughout the period of cardiopulmonary bypass. When the left atrial appendage could not be shown by transoesophageal echocardiography, the diagnosis of inverted left atrial appendage was finally entertained and then confirmed by external inspection of the heart. The inverted left atrial appendage was gently everted manually using forceps. After eversion of the left atrial appendage to its normal anatomical position, the "mass" inside the left atrium was no longer apparent on the second post-operative transoesophageal echocardiogram.

The patient was uneventfully extubated within 14 hours post-operatively. Unfortunately, without any important preceding or concomitant clinical events, the patient developed sudden asystole on post-operative day 2. She was resuscitated both medically and mechanically, but no recovery was achieved. The exact cause of death is not known.

Discussion

Inverted left atrial appendage is an unusual complication associated with surgery for congenital heart disease. It is often clinically asymptomatic and is most frequently detected during the post-operative echocardiogram.[1–7] During cardiac surgical procedures, this event can develop spontaneously or iatrogenically due to manipulation of the cardiac chambers. An inverted left atrial appendage can mimic an intracardiac mass in the left atrial cavity.[4] On echocardiography, a thumb-like mass with a broad base and hinge-type motion towards the mitral valve orifice are characteristic.[2] The differential diagnosis must include the presence of a thrombus, vegetation, or cardiac tumour within the left atrium.

There are several possible mechanisms for the occurrence of inversion of the left atrial appendage as a result of cardiac surgery.[6] This phenomenon can develop spontaneously or iatrogenically during surgical procedures. During cardiopulmonary bypass, the heart is decompressed using specialised vents. Negative suction is carefully applied through these vents during bypass; however, the left atrial appendage could theoretically invert as a result of this negative pressure. This event may also occur during placement or removal of these vents. This is especially true during deairing manoeuvres, which may include digital inversion of the left atrial appendage. Moreover, anatomical characteristics of the left atrial appendage may be a predisposing factor. The presence of a long, thin atrial appendage with a narrow base may be a potential risk for inversion of the left atrial appendage. For example, it has been noted that an inverted left atrial appendage can evert spontaneously while the heart is being filled.[6] Theoretically, the appendage may be less likely to evert if its base is narrow. Therefore, the surgeon should always confirm that the left atrial appendage is not inverted at the end of the surgical procedure.

Transoesophageal echocardiography is a useful technique to detect masses in the cardiac chambers during the intraoperative and post-operative periods. If intraoperative transoesophageal echocardiography is not available, an inverted left atrial appendage may go unnoticed.[1] In addition to transthoracic echocardiography, MRI has been used in the post-operative setting to diagnose inverted left atrial appendage.[3] Surgically confirmed inverted left atrial appendage cases have also been reported in the literature.[1–3] Spontaneous eversion of an inverted left atrial appendage has been documented while the heart is being filled and, in one case, within 1 year of surgery.[4,8]

In the literature, there are no inverted left atrial appendage cases associated with post-operative mortality. However, it has been reported that inverted left atrial appendage can develop spontaneously after cardiac surgery and this may lead to mitral valve

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obstruction.[6] The diagnosis of this event during the post-operative period should be considered an emergency situation, which necessitates prompt surgical intervention. When detected, the inverted left atrial appendage can be manually everted, similar to that in our case, or, in addition, it can also be ligated at its base to prevent future inversion.[1–3,6,8] In our case, the left atrial appendage was everted immediately after its diagnosis on transoesophageal echocardiography, but it was not ligated. Unfortunately, the patient died 2 days after the operation and the reason for cardiac arrest was unclear.

The cause of sudden mortality in this patient remains unexplained. She was uneventfully extubated 14 hours post-operatively. The sudden cardiac arrest developed on post-operative day 2 without any important preceding or associated clinical events, and no cardiac or pulmonary complication was observed before this event. The post-operative rhythm leading up to the acute event was normal sinus at 130 beats/minute without any evidence of heart block. The blood pressure immediately preceding the event was recorded as 82/54 mmHg with a central venous pressure of 10 mmHg and with a continuous arterial saturation of 100%. There was no evidence of increased work of breathing or stridor and the post-operative chest X-ray was normal. The chest tube drainage was 50 cc and the urine output was 142 cc over the 8 hours preceding the event. The arterial blood gas 1 hour before the acute event was normal with a lactate of 1.2 mmol/L and a base surplus of 4.6 mmol/L.

In congenital cardiac surgery, sudden events associated with mortality can develop because of fatal arrhythmias, pneumothorax, massive bleeding, or cardiac tamponade, none of which were present in our patient. In some patients, sudden airway obstruction or laryngospasm may be associated with fatal events. In this case, clinical follow-up was uneventful until the sudden cardiac arrest developed. Given the lack of other obvious and more common causes for the sudden clinical deterioration, spontaneous and recurrent left atrial inversion may be a possible explanation for the sudden and unheralded mortality in this patient.

In conclusion, increased awareness on the possibility of developing an inverted left atrial appendage will lead to a more prompt diagnosis. The intraoperative or post-operative diagnosis of this entity requires prompt intervention by manual reduction of the left atrial appendage with external or internal ligation of its base to prevent recurrence. The paediatric cardiac surgeon should routinely examine the heart externally, after decannulation and before sternal closure, to rule out inverted left atrial appendage.

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Conflicts of Interest

None.

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