Cardiology in the Young

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Right atrial appendage aneurysm: an updated systematic review

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Review

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Abstract

Background: Right atrial appendage aneurysm, or giant right atrial appendage, is extremely rare, with very few cases reported in scientific literature. We sought to systematically review the published cases of right atrial appendage aneurysm in terms of age, sex, clinical presentation, electrocardiography, imaging (chest X-ray, echocardiography, CT/cardiac magnetic resonance), and outcome. *Methodology:* An electronic search for case reports, case series, and related articles published until March 2025 was carried out, and clinical data were extracted and analysed. *Results:* Forty-four cases of right atrial appendage aneurysm were identified with a clear male prevalence (68.2%) and commonly presenting in the third decade of life. Palpitation (27.3%) and dyspnoea (18.2%) were the most common clinical presentations, whereas 40.9% of right atrial appendage aneurysm patients were asymptomatic. Electrocardiography was done in 77.3% of the sample. It displayed an atrial arrhythmia (atrial fibrillation or flutter, atrial tachycardia, supraventricular tachycardia) in 31.8%. A chest X-ray was done in 65.9%. Echocardiography was the most common diagnostic modality (93.2%). Right atrial appendage aneurysm diagnosis was confirmed on CT and/or MRI in 79.5%. The mean size of the right atrial appendage aneurysm was 93 × 70 mm. In 12 patients (27.3%), an associated congenital cardiac abnormality was found, mostly in the form of an atrial septal defect/patent foramen ovale (22.7%). Half of the patients (50.0%) were treated surgically, whilst 47.8% were treated medically with close follow-up. One patient experienced right atrial appendage aneurysm reduction in size after atrial septal defect device closure. One death (2.3%) was reported also. *Conclusion:* Although very uncommon, right atrial appendage aneurysm can be linked to considerable morbidity. Surgical removal is recommended for patients who are symptomatic.

Introduction

Right atrial appendage aneurysms or giant right atrial appendages are uncommon occurrences. They are characterised by an abnormal dilation or outpouching of the right atrial appendage. This structural anomaly may result from congenital weakness of the atrial wall or secondary factors such as increased right atrial pressure.[1] There are congenital as well as acquired cases, but, unlike left atrial appendage aneurysm, a formal definition of right atrial appendage aneurysm is currently lacking.[2] Right atrial appendage aneurysms are less frequent than those of the left atrial appendage.[1] This condition can affect individuals of all ages and is frequently discovered incidentally during cardiac imaging done for other purposes. These aneurysms may be asymptomatic or cause symptoms.[1]

So far, the first and only systematic review on right atrial appendage aneurysm dates back to 2014, encompassing just 17 cases published between 2001 and July 2013.[1] The present review aims at updating the list of the analysed right atrial appendage aneurysm cases. While the earlier review captured the state of knowledge on right atrial appendage aneurysm with remarkable clarity, subsequent advances necessitate a reassessment of its conclusions. In fact, many additional cases published in recent years are likely to have expanded the understanding of the clinical spectrum, potential complications, and therapeutic options for right atrial appendage aneurysm.

Search methodology and data gathering

An independent electronic search of Medline, PubMed, and Scopus was carried out by two authors (PPB and KM) for case reports on right atrial appendage aneurysm published up until March 2025. The search utilised terms such as "right atrium," "appendage," "aneurysm,"

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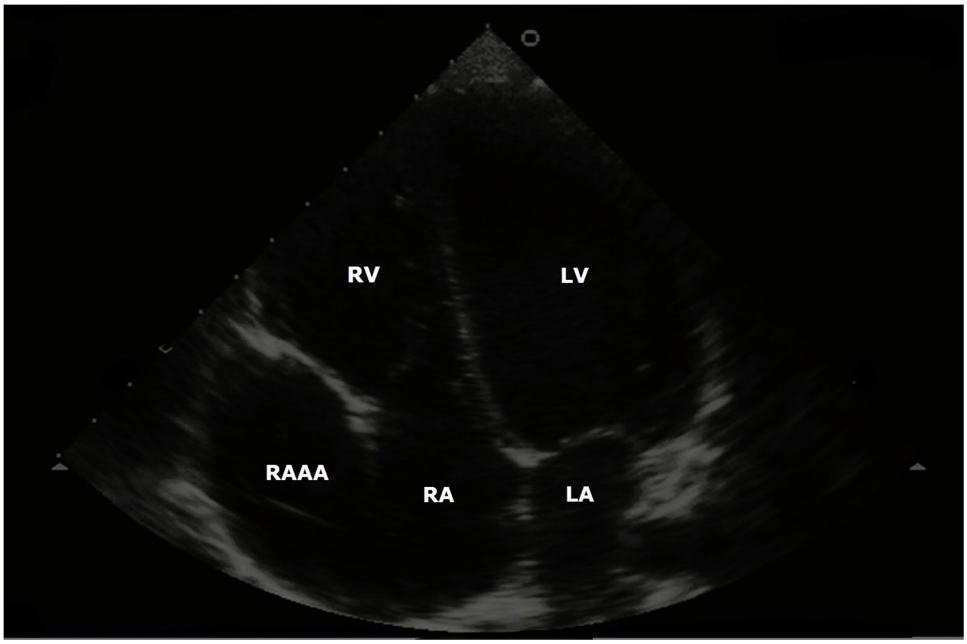


Figure 1. Echocardiographic 4 chambers view displaying a right atrial appendage aneurysm. Acronyms: RA = Right Atrium; RV = Right Ventricle; LA = Left Atrium; LV = Left Ventricle.

"cardiac," "congenital," "atrial," and "acquired," along with the medical subject heading terms "atrial appendage" and "aneurysm." The search was restricted to human studies.

Cases that only described right atrial aneurysm were excluded from the study. No restrictions based on time or language were applied. The references of the reviewed articles were manually searched to find additional cases, with efforts made to prevent duplication. A total of forty-four cases were identified (see Figure 1 for the PRISMA diagram of the paper selection process). Various factors, including demographic information, clinical presentations, electrocardiographic features, diagnostic methods, treatment options, and outcomes, were analysed.

Results

A total of 44 cases of right atrial appendage aneurysm were identified, comprising 30 males (68.2%) and 13 females (29.5%) (see Table 1).[3–46] In one case sex was not specified.[12] The median age at presentation was 27.3 ± 24.4 years, with an age range between 0 and 72 years. The highest right atrial appendage aneurysm prevalence occurred in the third decade of life. Eighteen patients (40.9%) were asymptomatic. The most frequent clinical symptom was palpitation, affecting 12 patients (27.3%), followed by dyspnoea in 8 patients (18.2%). Other symptoms were seen in 13.6% of the sample. Electrocardiography was documented for 34 patients (77.3%), of whom 14 (31.8%) had an atrial arrhythmia (atrial fibrillation or flutter, atrial tachycardia, supraventricular tachycardia). Normal sinus rhythm was detected in 19 subjects (43.2%). Chest radiography was done in 29/44 (65.9%) and revealed cardiomegaly in 20 (45.5%). Echocardiography was the most common diagnostic modality (93.2%). Right atrial appendage aneurysm diagnosis was confirmed on CT and/or MRI in 79.5%. These imaging studies were conducted to further assess and confirm the diagnosis of right atrial appendage aneurysm identified through echocardiographic examinations. Right atrial appendage aneurysm dimensions were measured in 19/44 (43.2%). The mean size of right atrial appendage aneurysm was 93 × 70 mm. In 12 patients (27.3%), an associated congenital cardiac abnormality was found, mostly in the form of an atrial septal defect/patent foramen ovale (22.7%). One patient presented with aneurysms in both the right and left atrial appendages, along with Eisenmenger's syndrome resulting from underlying atrial and ventricular septal defects.[40] Half of the patients (50.0%) were treated surgically with or without the Maze procedure (performed in 4.5%), whilst 47.8% were treated medically with close follow-up. The indications for surgery were not clearly mentioned in some cases. Medical treatment included antiarrhythmic drugs and anticoagulant therapy against thromboembolic risk. In a few case reports the outcome was not reported. One death (2.3%) was reported also (see Table 2).

Discussion

Right atrial appendage aneurysm is an uncommon condition which can be either congenital or acquired.[1] In our review of 44 documented cases, males were affected way more often than females.[1,47] Although right atrial appendage aneurysm has been observed across all age groups, it is most commonly diagnosed in individuals in their 30s. It has also been identified in newborns and even during prenatal development.

The congenital form of right atrial appendage aneurysm is thought to result from abnormal development of the right atrial muscular wall.[30] The exact cause of the acquired form remains unclear, but it is suspected to be associated with prolonged elevation of right atrial pressure and increased atrial volume due to underlying conditions such as pulmonary hypertension, congenital heart defects like atrial septal defect, or heart failure.[1]

The coexistence of left atrial appendage aneurysm and patients with right atrial appendage aneurysm is anecdotal.[40] Histological examination often reveals endocardial fibrosis, and in some cases, lipomatous degeneration of the aneurysm is observed without signs of inflammation.[19,44]

The most frequent symptom of right atrial appendage aneurysm was palpitation caused by atrial tachyarrhythmias, such as atrial fibrillation/flutter, atrial tachycardia, or supraventricular tachycardia. Some individuals with right atrial appendage

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Table 1. List of the case reports which have been published in the field

Author	Patient´s age at diagnosis (years)	Gender	Clinical presentation	ECG	Chest X-ray	Diagnosis by echocardiography	Diagnosis by CT/MRI	Associated cardiac abnormalities	Outcome
Luo et al. [3]	2	Male	Asymptomatic	Sinus rhythm	Yes	No	Yes	None	Successful surgery
Yue et al. [4]	1 (initial prenatal suspicion at 26 weeks gestation)	Male	Dyspnoea, arrhythmia	Supraventricular tachycardia	Yes	Yes	No	Patent foramen ovale	Successful surgery
Niu et al. [5]	57	Female	Congestive heart failure	Sinus rhythm	Yes	Yes	No	None	Successful surgery
Hiremath et al. [6]	37	Male	Palpitations	Atrial Flutter	Yes	Yes	No	None	Successful surgery
Uzun et al. [7]	33	Male	Asymptomatic	Sinus rhythm	Yes	Yes	No	None	Conservative management
Hiraoka et al.[8]	59	Female	Asymptomatic	No	Yes	Yes	No	None	Successful surgery
Zhang et al.[9]	4 years and 7 months old	Male	Arrhythmia, palpitations	Atrial flutter / Atrial fibrillation	No	Yes	No	None	Successful surgery
Sivakumaran et al. [10]	Elderly patient (50–70 years, exact age not disclosed)	Male	Asymptomatic	No	No	Yes	No	None	Conservative management
Celik et al. [11]	22	Male	Palpitations	Sinus rhythm	No	Yes	No	None	Conservative management
Tunks et al.[12]	Prenatal (detected at 28 weeks gestation), confirmed postnatally	Not mentioned	Asymptomatic	Sinus rhythm	No	Yes	No	Small secundum atrial septal defect	Successful surgery
Fang et al. [13]	44	Male	Palpitations	Atrial fibrillation	No	Yes	No	None	Successful surgery
Qian et al. [14]	36	Male	Exertional fatigue	No	No	Yes	No	None	Successful surgery
Toufan et al. [15]	65	Male	Palpitations, congestive heart failure	Atrial flutter	No	Yes	No	None	Conservative management
Sondhi et al. [16]	22	Female	Asymptomatic	No	Yes	Yes	No	None	Successful surgery
Cianciulli et al. [17]	57	Male	Asymptomatic	Atrial fibrillation	No	Yes	No	None	Surgery refused, conservative treatment
Contreras et al. [18]	52	Male	Congestive heart failure	Sinus rhythm	Yes	Yes	No	None	Conservative management
Guaricci et al. [19]	39	Female	Palpitations	Supraventricular tachycardia	Yes	Yes	No	Patent foramen ovale	Successful surgery
Wang et al. [20]	1	Male	Palpitations	Supraventricular tachycardia	Yes	Yes	No	None	Successful surgery
Nam et al.[21]	51	Male	Asymptomatic	Sinus rhythm	Yes	Yes	No	None	Conservative management
Miranda et al.[22]	Prenatal diagnosis at 25 weeks of gestation	Male	Asymptomatic	No	No	Yes	No	Patent foramen ovale	Conservative management

(*Continued*)

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Table 1. (*Continued*)

Author	Patient´s age at diagnosis (years)	Gender	Clinical presentation	ECG	Chest X-ray	Diagnosis by echocardiography	Diagnosis by CT/MRI	Associated cardiac abnormalities	Outcome
Zhou R, et al. [23]	7	Male	Heart murmur assessment	Sinus rhythm	Yes	Yes	No	None	Successful surgery
Belegrinos et al. [24]	10 Months old	Male	Heart murmur assessment	Sinus rhythm	Yes	Yes	No	None	Initially: Conservative management. At age 10: successful surgery
Vidiyala et al.[25]	2	Male	Cyanosis and failure to thrive	No	No	No	Yes	Double outlet right ventricle. Ventricular septal defect. Pulmonary stenosis. Juxtaposed atrial appendages. Anomalous systemic venous drainage	Conservative management
Narumi et al.[26]	35	Male	Palpitations	Supraventricular tachycardia	No	No	No	None	Conservative management
Hafez et al.[27]	6 months	Male	Dyspnoea, palpitations	No	Yes	Yes	No	None	Successful surgery
Duan et al.[28]	13	Female	Chest Pain, dyspnoea, heart murmur assessment	Sinus rhythm	No	Yes	No	Atrial septal defect	Transcatheter closure of ASD. RAAA: conservative management
Tejero-Hernández et al. [29]	2 days (prenatal suspicion; confirmed postnatally)	Female	Asymptomatic	Sinus rhythm	Yes	No	Yes	Patent foramen ovale	Conservative management
Ishii et al. [30]	38 weeks of gestation (prenatal diagnosis); confirmed postnatally	Female	Asymptomatic	Sinus rhythm	Yes	Yes	No	None	Conservative management
Nakahara et al.[31]	68	Male	Palpitations	Atrial fibrillation	Yes	No	No. Intraoperative finding: Also small RAA aneurysm undetected on imaging	None	Successful surgery
Gulati et al.[32]	13	Male	Exertional breathlessness, heart murmur assessment	Sinus rhythm	Yes	No	Yes	None	Initially conservatively. Surgical intervention after aneurysm growth and RV compression
Yue et al. [33]	Approximately 41 years (RAAA detected 10 years before current age of 51)	Female	Asymptomatic	No	No	Yes	No	None	Conservative management
Sate et al. [34]	53	Male	Acute ST-elevation myocardial infarction (STEMI). Dextrocardia discovered on physical examination	STEMI	Yes	Yes	No	Dextrocardia. Anomalous origin of right coronary artery (RCA) from left coronary sinus. Two-vessel coronary artery disease	Successful surgery

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Table 1. (*Continued*)

Faqeeh et al.[35]	1 day old (newborn)	Female	Jaundice, heart murmur assessment	No	Yes	Yes	No	Atrial septal defect, ventricular septal defects, patent ductus arteriosus	Conservative management
Bornaun et al. [36]	Prenatal diagnosis at 21 weeks gestation; confirmed postnatally	Male	Asymptomatic	Sinus rhythm	Yes	Yes	No	None	Conservative management
Xu et al. [37]	62	Male	Chest pain	SR	No	Yes	No	None	Conservative management
Le Ven et al.[38]	16	Male	Heart murmur assessment	SR	Yes	No	Yes	None	Conservative management
Duran et al.[39]	72	Male	Claudication	Sinus rhythm	Normal	Yes	No	None	Underwent abdominal aorta grafting and aortobifemoral bypass because of abdominal aortic aneurysm . Discharged on oral anticoagulant therapy. No post-op complications reported
Sevimli et al.[40]	22	Male	Palpitations, dyspnoea, cyanosis, clubbing of fingers and toes	Multifocal atrial tachycardia	Yes	Yes	No	Concomitant left atrial appendage aneurysm. Large atrial septal defect. Large muscular ventricular septal defect. Severe pulmonary hypertension (pulmonary artery systolic pressure: 130 mmHg). Eisenmenger's syndrome	Inoperable due to Eisenmenger syndrome. Conservative management
Barberato et al. [41]	23	Male	Palpitations	Atrial flutter	Yes	Yes	No	Patent foramen ovale	Conservative management
Mizui et al.[42]	11 days old (tachycardia noted at birth; surgery at 11 months)	Female	Palpitations, congestive heart failure	Ectopic atrial tachycardia	Yes	No. Aneurysm seen on angiography post-ablation	No	Atrial septal defect	Successful surgery
Dalvi et al.[43]	22	Male	Congestive heart failure	Sinus rhythm	Yes	Yes	No	None	Successful surgery
Corti et al. [44]	20	Female	Arrhythmia	Atrial tachycardia	Yes	No	Yes	None	Successful surgery
Kurata et al.[45]	18	Female	–	–	–	–	–	–	Death (Autoptic diagnosis)
Yoon et al.[46]	69	Female	Chest pain	Sinus rhythm	Yes	No	Yes	None	Conservative management

Table 2. Right atrial appendage aneurysm patients' features

Male-to-female ratio	2.3/1
Mean age at diagnosis	27.3 ± 24.4 years (range 0–72
Symptoms	Asymptomatic (40.9%)
	Palpitation (27.3%)
	Dyspnoea (18.2%)
ECG changes	Atrial fibrillation/flutter/atrial or supraventricular tachycardia (31.8%)
Imaging	Echocardiography (93.2%)
	CT/MRI (79.5%)
Associated congenital heart disease	27.30%
Surgery	50.00%
Conservative management	47.80%
Death	2.30%

diagnosis. In cases where transthoracic images were inconclusive, transoesophageal echocardiography with or without contrast enhancement was employed.[13,19] Advanced imaging techniques such as CT and MRI were valuable adjuncts, offering high sensitivity and specificity. These modalities were particularly useful for confirming the connection of the aneurysm to the right atrial appendage and assessing its relationship to adjacent structures.[16]

Surgical resection is recommended for symptomatic patients and those with enlarging right atrial appendage aneurysms to reduce the risk of arrhythmias and thromboembolic events. Concomitant procedures, such as bi-atrial aneurysm resections, the Maze procedure, and CHD repair, can be performed simultaneously to address associated cardiac conditions.[18,19,40,42] Currently, there is no information available regarding the use of device direct closure for right atrial appendage aneurysm. While device-based closures are commonly employed for left atrial appendage management to reduce stroke risk in atrial fibrillation patients,[48] their application in right atrial appendage aneurysm cases is not documented yet. One patient experienced right atrial appendage aneurysm reduction in size after atrial septal defect

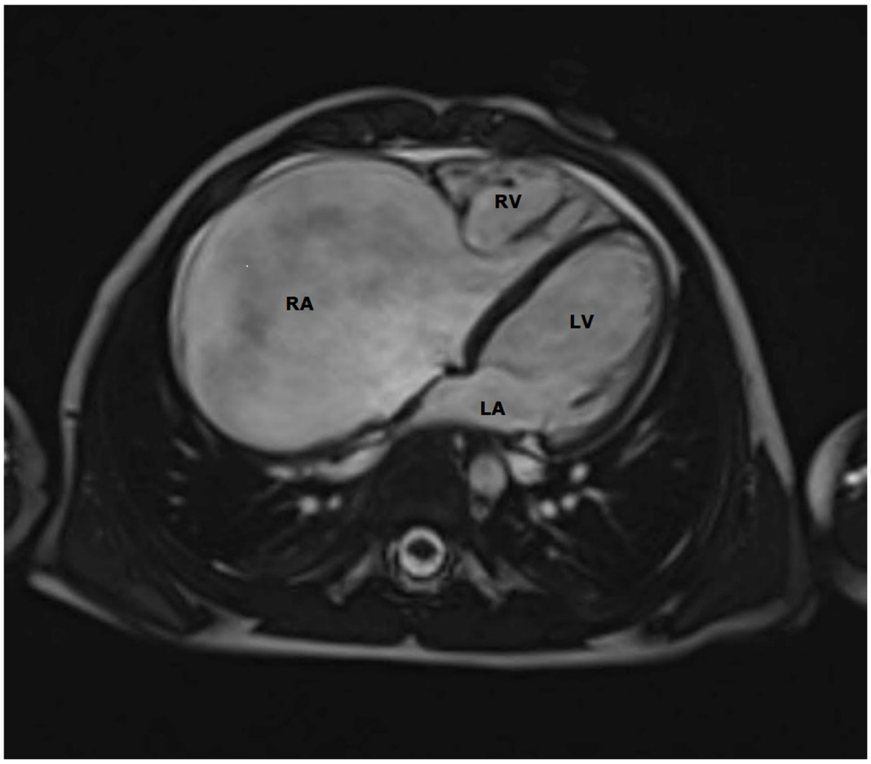


Figure 2. Axial four chamber view on cardiac MRI showing a massive right atrial dilatation in a 10-month-old patient (unpublished case). Acronyms: RA = Right Atrium; RV = Right Ventricle; LA = Left Atrium; LV = Left Ventricle.

aneurysm exhibited no symptoms, with the condition being discovered incidentally during cardiac imaging, autopsy, or surgeries conducted for unrelated reasons.[45] Others presented with thromboembolic complications.[29] Dyspnoea, often linked to underlying cardiovascular issues such as heart failure and pulmonary hypertension, can also be a symptom. In our review, some patients had pulmonary hypertension with or without Eisenmenger's syndrome or had left ventricular dysfunction.[40–42]

Cardiomegaly, indicative of underlying heart disease, was observed on chest X-rays in 45.5% of patients with right atrial appendage aneurysm. Transthoracic echocardiography served as the initial imaging modality for right atrial appendage aneurysm

device closure.[28] Surgical resection remains the standard treatment for symptomatic or enlarging right atrial appendage aneurysms to mitigate risks such as arrhythmias and thromboembolic events.[1]

Less than half of right atrial appendage aneurysm patients were managed conservatively without reported complications. A conservative approach is typically recommended for asymptomatic individuals.[1] However, factors such as large right atrial appendage aneurysm size, enlarged right atrium, and associated congenital heart defects have been linked to increased risks of thromboembolic events and arrhythmias.[18,19,40,42] Regular monitoring of asymptomatic patients using CT imaging or cardiac magnetic resonance to detect right atrial appendage aneurysm

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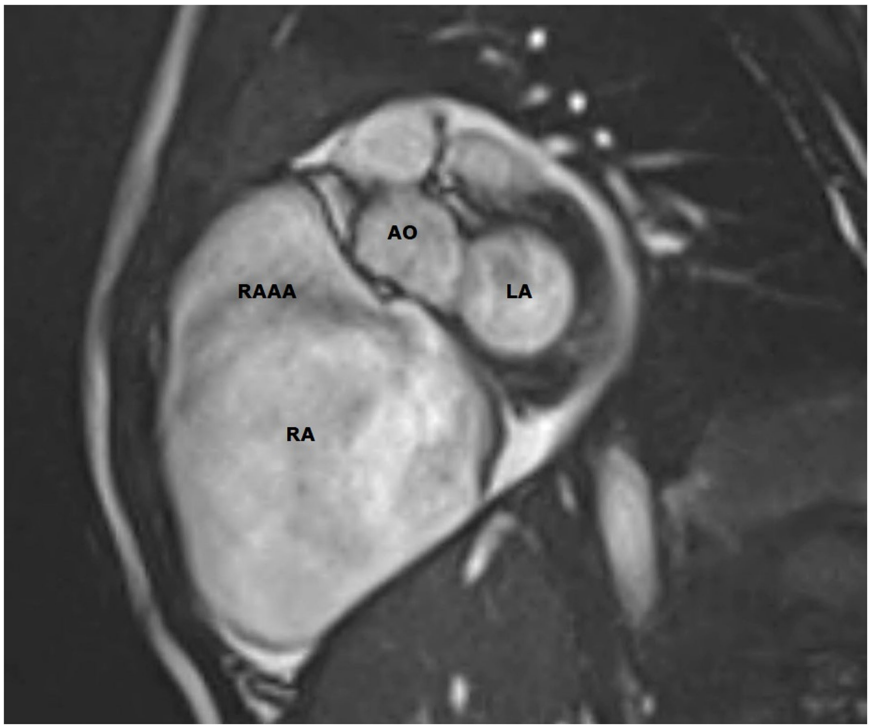


Figure 3. Cardiac MRI (short axis view) displaying the presence of a right atrial appendage aneurysm in the same baby who has subsequently undergone surgery for aneurysm plication. Acronyms: RAAA = Right Atrial Appendage Aneurysm; RA = Right Atrium; AO = Aorta; LA = Left Atrium.

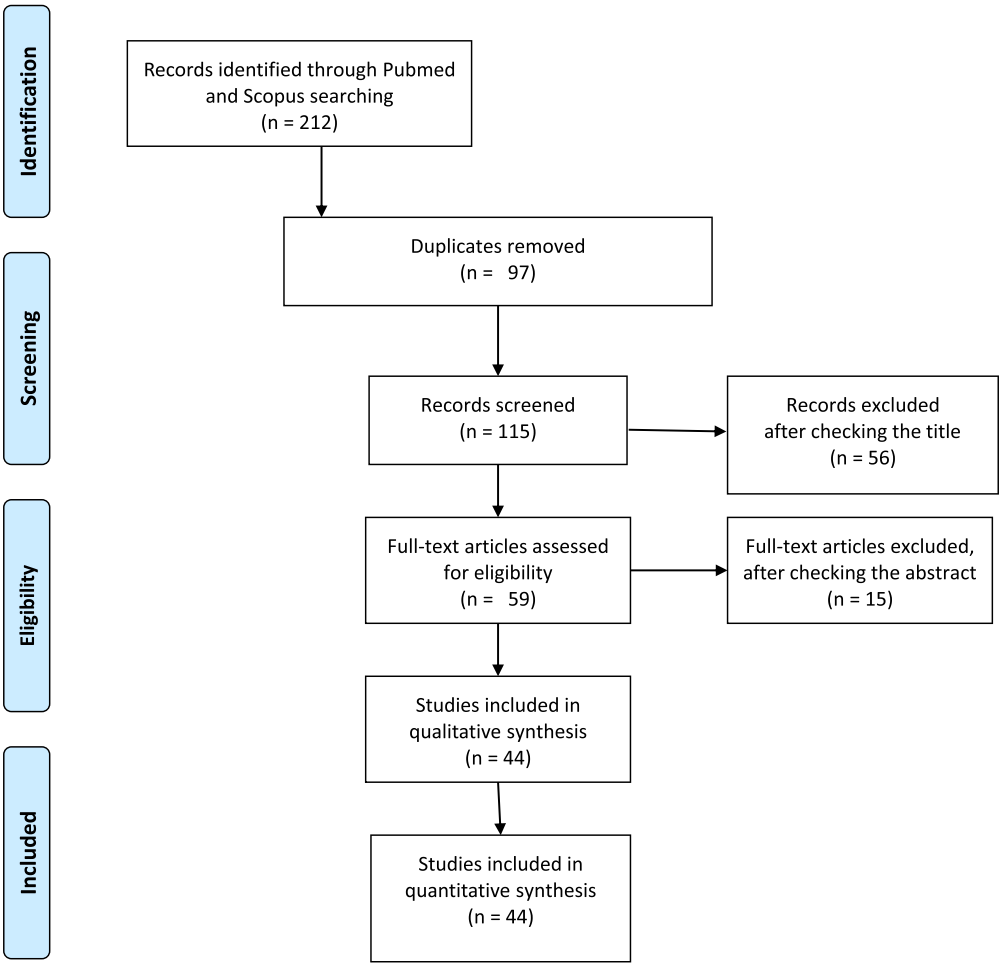


Figure 4. Prisma flow diagram.

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enlargement has proven useful in identifying candidates who may benefit from early surgical intervention.[32]

Asymptomatic right atrial appendage aneurysm patients undergoing conservative management should receive anticoagulation therapy to prevent thrombus formation. Antiarrhythmic medications are used to manage atrial tachyarrhythmias. The prognosis for these patients is generally favourable, but they require close monitoring.[17]

A large or progressively enlarging right atrial appendage aneurysm has been identified as a risk factor for thromboembolic complications and atrial arrhythmias. Although there is limited long-term data supporting routine repeat imaging, some experts suggest follow-up cardiac MRI every two years.[32]

Patients with evidence of aneurysm enlargement should be considered for surgical excision due to their heightened risk of arrhythmias and thromboembolic events. Those with a patent foramen ovale or atrial septal defect should be carefully monitored for systemic thromboembolism, and these defects should be closed during right atrial appendage aneurysm surgical treatment.[30,49]

Potential complications of right atrial appendage aneurysm include pulmonary embolism, rupture of the aneurysmal right atrial appendage, and sudden death.[17]

In conclusion, right atrial appendage aneurysm can lead to significant health issues. Surgical removal is advised for symptomatic individuals and those with large aneurysms. Asymptomatic patients may be managed conservatively, focusing on preventing thromboembolic events and atrial tachyarrhythmias. Regular follow-up imaging, such as echocardiography and cardiac magnetic resonance, can be considered to monitor aneurysm size and progression (see Figures 2, 3, 4).

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