


# Blood cysts of the cardiac valves in adults: Review and analysis of published cases

Uberto Bortolotti MD<sup>1</sup>  | Igor Vendramin MD<sup>1</sup> | Andrea Lechiancole MD<sup>1</sup> | Sandro Sponga MD<sup>1</sup> | Angela Pucci MD<sup>2</sup> | Aldo D. Milano MD, PhD<sup>3</sup> | Ugolino Livi MD<sup>1</sup>

<sup>1</sup>Cardiothoracic Department, University Hospital of Udine, Udine, Italy

<sup>2</sup>Division of Pathology, Pisa University Hospital, Pisa, Italy

<sup>3</sup>Division of Cardiac Surgery, University of Bari Aldo Moro, Bari, Italy

## Correspondence

Uberto Bortolotti, MD, Largo Traiano 23, Montegrotto Terme (PD), Italy.  
Email: uberto48@gmail.com

## Abstract

**Background and aim:** Blood cysts of cardiac valves are generally seen in newborns and infants and very rarely in adults. Although in most cases they are incidental findings they may be associated to severe cardiac or systemic complications. This study analyzes incidence, presentation, and treatment of valvular blood cysts in adults.

**Methods:** A review of the pertinent literature through a search mainly on PubMed and Medline was performed.

**Results:** In patients  $\geq 18$  years of age, our search disclosed 54 patients with mitral blood cysts (mean age,  $48 \pm 18$  years), 9 with a tricuspid valve cyst (mean age,  $67 \pm 15$  years), 3 with a blood cyst on the pulmonary valve (age 31, 43, and 44 years), and 1 aortic valve cyst in a 22-year-old man. Most patients were asymptomatic while stroke, syncope, or myocardial infarction occurred in six patients with a mitral valve cyst. Blood cysts were removed surgically in 70% of patients with a mitral cyst, in 55% with a tricuspid cyst, and in all those with a pulmonary or aortic cyst. At histology, the cyst wall was composed mainly by fibrous tissue and with the inner surface lined with typical endothelium.

**Conclusions:** Blood cysts of cardiac valves are rare in adults but may cause life-threatening complications, particularly when located on the mitral valve. For such reason, surgical removal appears advisable, with low-risk procedures. Widespread use of multimodality imaging techniques will most likely increase the number of valvular blood cysts diagnosed also in adults.

## KEYWORDS

cardiovascular pathology, valve repair/replacement

Uberto Bortolotti and Igor Vendramin contributed equally to this work.

This is an open access article under the terms of the Creative Commons Attribution License, which permits use, distribution and reproduction in any medium, provided the original work is properly cited.

© 2021 The Authors. *Journal of Cardiac Surgery* published by Wiley Periodicals LLC.

## 1 | INTRODUCTION

Benign cardiac neoplasms are substantially rare and among these intracardiac cysts filled with blood are even more uncommon; indeed, they have not even been mentioned in a recent review of cardiac tumors reported by Tyebally et al.<sup>1</sup> However, mainly owing to advancement in diagnostic techniques, with a widespread use of and accessibility to multimodality imaging, an increase in the diagnosis of cardiac or pericardial masses has been observed recently and is expected to further grow in the future.<sup>1</sup> Intracardiac blood cysts (BCs) are commonly found in newborns and infants being rare after the first year of life, they mainly involve the cardiac valves and generally disappear during growth.<sup>2</sup> In adults they may occasionally be diagnosed, are usually located on all cardiac valves and chambers, the mitral valve (MV) being the most frequently involved; despite absence of histological malignancy BCs of the heart valves have been reported to cause life-threatening complications with even severe sequelae.

There is still uncertainty on the origin of BCs and controversy on whether medical treatment with continuous clinical and echocardiographic patient follow-up should be preferred to immediate surgical removal. The present review, analyzing the published cases, was undertaken with the aim of assessing the incidence, evaluating the clinical presentations, and discussing the treatment of BCs involving the cardiac valves in adults.

## 2 | METHODS

We have performed a search of the English literature, through PubMed and Medline, to identify all cases of BCs involving the MV, MV apparatus, and tricuspid, aortic and pulmonary valves (AV and PV) that have been so far reported. Articles in textbooks and meeting abstracts were excluded, as well as cases included in general reviews of cardiac tumors or clinical reports, when detailed clinical and pathological information on BCs, when present, was lacking. Cases of valvular cysts but with the uncertain histological diagnosis were excluded. The reference sections of pertinent articles were also evaluated as well as personal files and the archives of journals available on the CTSNet website. All reported cases observed in the pediatric population were excluded, considering only those occurring in patients  $\geq 18$  years of age.

Main terms used in the literature search, alone or in combination, were intracardiac cyst, valvular cyst, BC, blood-filled cyst, pedunculated cysts, MV, MV apparatus, MV leaflet, tricuspid valve (TV), papillary muscle, chordae tendineae, AV, PV, semilunar valve, valve cusps, atrio-ventricular valve, and semilunar valve.

Approval by the Ethical Committees was not required for this kind of study, as well as informed patient consent, provided that all relative data were treated anonymously.

## 3 | RESULTS

### 3.1 | BCs of the MV

Our literature search has documented a total of 50 articles considered eligible for analysis and reporting a total of 54 patients; 36 (72%) are reports of a single case,<sup>3–38</sup> 12 (24%) images in cardiovascular disease,<sup>39–50</sup> 1 case series reporting 5 patients (2%),<sup>51</sup> and 1 letter to the editor (2%),<sup>52</sup> all published from 1960 to 2021 (Tables 1 and 2).

#### 3.1.1 | Patient characteristics

There were 29 males (55%) and 25 females (45%) with an age ranging from 18 to 87 years (mean,  $48 \pm 18$  years). The majority of patients was asymptomatic and the cyst was disclosed at routine controls for other pathologies. Symptoms related to the presence of MV BCs, in the absence of any other intracardiac abnormality, included predominantly palpitations, chest discomfort, and exertional dyspnea. In three patients, BCs caused severe left ventricular outflow tract (LVOT) obstruction,<sup>5,11,50</sup> two patients presented with hemiparesis,<sup>4,43</sup> two with syncopal episodes,<sup>32,50</sup> two had a stroke,<sup>15,18</sup> two a myocardial infarction,<sup>9,21</sup> and in one the BC caused severe MV regurgitation.<sup>23</sup> Except for the first two BCs cases described, diagnosed either by cineangiography<sup>3</sup> or M-mode echocardiogram,<sup>5</sup> in all other cases transthoracic 2D echo was used for evidencing the presence of an MV mass (Figure 1). The diagnosis was confirmed by supplemental transesophageal echo in many cases and more recently by multimodality imaging, including angio-computed tomography, nuclear magnetic resonance, contrast real-time echocardiography, and 3D echo.<sup>23,34,36,38,44,45,49,49,51</sup>

#### 3.1.2 | Surgical data

Cyst removal was performed in 37 patients (70%) while in 16 (30%) medical treatment and serial follow-up was recommended; in one patient AV and ascending aorta replacement were performed leaving in place a small BC on the MV.<sup>20</sup> Surgery was generally performed through a standard median sternotomy while in three cases a minimally invasive, endoscopic technique was used.<sup>27,36,45</sup> Two patients had previous open heart operations and required repeat sternotomies,<sup>22,42</sup> while one had a previous liver transplantation.<sup>10</sup> The BC was approached through a trans-aortic incision in 7 cases, through the left atrium in 6, trans-septal in 4, while in 20 cases it was not specified. Beside cyst removal, 27 patients (73%) had associated procedures: mitral valve replacement (MVR) in 12, mitral valve repair (MVR) in 9, coronary artery bypass grafting in 3, and combined MVR and AV replacement in 1. In addition, in two patients, with associated *cor triatriatum*, resection of the intraatrial band was performed.<sup>14,24</sup> No operative deaths are reported.

**TABLE 1** Characteristics of case reports and case series of blood cysts of the mitral valve apparatus

Author, ref. no.	Year	Age, sex	Location	Max. dimension, mm	Surgical approach	Operation
Leatherman et al. <sup>3</sup>	1968	39, F	AML	30	Transaortic	Cyst removal
Hauser et al. <sup>4</sup>	1983	27, M	APM	25	LA incision	Cyst removal, MVR
Arnold et al. <sup>5</sup>	1990	46, M	AML	32	Transaortic	Cyst removal, MVR
Xie et al. <sup>6</sup>	1992	41, F	PML	13, 3 <sup>a</sup>	Transseptal	Cysts removal
Ohmoto et al. <sup>7</sup>	1993	57, M	APM	40	Transaortic	Cyst removal
Pelikan et al. <sup>8</sup>	1999	50, M	AML	22	No surgery	-
Sharma et al. <sup>9</sup>	2000	68, M	APM	20	Transaortic	Cyst removal, CABG
Kuvin J et al. <sup>10</sup>	2004	45, F	AML	25	LA incision	Cyst removal
Minnecci et al. <sup>11</sup>	2004	44, F	AML	20	Transaortic	Cyst removal, MVR
López-Pardo et al. <sup>12</sup>	4008	34, M	AML	22	No surgery	-
Tsutsui et al. <sup>13</sup>	2008	47, F	AML	15	No surgery	-
Denker et al. <sup>14</sup>	2009	65, F	AML, PML	10, 10 <sup>a</sup>	NA	Cysts removal, MVr, LA band resection <sup>b</sup>
Lodha et al. <sup>15</sup>	2009	74, F	AML	NA	No surgery	.
Park et al. <sup>16</sup>	2009	22, M	APM	21	NA	Cyst removal, MVr
Migliore et al. <sup>17</sup>	2010	18, F	AML	22	No surgery	-
Khan et al. <sup>18</sup>	2012	87, F	AML	17	No surgery	-
Park et al. <sup>19</sup>	2012	47, M	APM	18	NA	Cyst removal, MVr
Bhatt et al. <sup>20</sup>	2013	45, F	PML	NA	-	No removal
Donndorf et al. <sup>21</sup>	2013	55, M	PM (n.s.)	20	Transaortic	Cyst removal, CABG
Ansari et al. <sup>22</sup>	2015	55, M	AML	26	LA incision	Cyst removal, CABG
Halim et al. <sup>23</sup>	2015	23, M	AML	20	Transseptal <sup>c</sup>	Cyst removal, MVr
Madhavan et al. <sup>24</sup>	2015	70, F	AML	16	NA	Cyst removal, MVR
Özmen et al. <sup>25</sup>	2015	19, F	AML	17	NA	Cysts removal, RA band resection <sup>b</sup>
Yilmaz et al. <sup>26</sup>	2015	63, M	AML	17	No surgery	-
Ahmad et al. <sup>27</sup>	2016	25, M	PPM	20	NA	Cyst removal, MVr
Okamoto et al. <sup>28</sup>	2016	41, F	PPM	29	LA incision	Cyst removal, MVR
Akutsu et al. <sup>29</sup>	2017	57, M	AML	15	Transseptal	Cyst removal, MVr
Pavsic et al. <sup>30</sup>	2017	44, F	AML	23	NA	Cyst removal, MVR
Bagheri et al. <sup>31</sup>	2018	62, M	PPM	21, 19 <sup>a</sup>	NA	Cysts removal, MVR
Ludhwani et al. <sup>32</sup>	2019	47, F	MVA	15	No surgery	-
Ma et al. <sup>33</sup>	2019	32, M	AML	46	NA	Cyst removal
Cerik et al. <sup>34</sup>	2020	42, F	AML	17	No surgery	-
Ramirez-Mesias et al. <sup>35</sup>	2020	57, F	AML	10	No surgery	-
Wang et al. <sup>51</sup>	2020	30, M	AML	17	NA	Cysts removal
		45, M	AML	25	NA	Cyst removal, MVR
		57, F	AML	11	NA	Cyst removal, MVR
		58, M	AML	19	NA	Cyst removal
		57, M	MV chordae	12	No surgery	-

(Continues)

TABLE 1 (Continued)

Author, ref. no.	Year	Age, sex	Location	Max. dimension, mm	Surgical approach	Operation
Beale et al. <sup>36</sup>	2021	62, F	AML	20	No surgery	-
Ionac et al. <sup>37</sup>	2021	39, M	AML	40	NA	Cyst removal, MVr
Zhang et al. <sup>38</sup>	2021	38, M	APM	25	LA incision	Cyst removal

Abbreviations: AML, anterior mitral leaflet; APM, anterior papillary muscle; CABG, coronary artery bypass grafting; F, female; LA, left atrium; M, male; MV, mitral valve; MVA, mitral valve annulus; MVr, mitral valve repair; MVR, mitral valve replacement; NA, not available; n.s., not specified; PM, papillary muscle; PML, posterior mitral leaflet; PPM, posterior papillary muscle; RA, right atrium.

<sup>a</sup>These patients had 2 cysts.

<sup>b</sup>These patient had a cor triatriatum sinister and dexter, respectively.

<sup>c</sup>This patient had a re sternotomy due to a previous closure of an atrial septal defect.

TABLE 2 Characteristics of cases reported as images or letter to the editor of blood cysts of the mitral valve apparatus

Author, ref. no.	Year	Age, sex	Location	Max. dimension, mm	Surgical approach	Operation
Kurtoğlu et al. <sup>39</sup>	2005	35, M	AML	21	No surgery	-
Yamamoto et al. <sup>40</sup>	2005	25, F	AML	25	NA	Cyst removal, MVr
Akinci and Celikyurt <sup>41</sup>	2010	64, M	AML	18	No surgery	PTCA
Combaret et al. <sup>42</sup>	2012	66, M	AML	16	NA	Cysts removal, MVR
Grimaldi et al. <sup>43</sup>	2012	63, F	AML	15	No surgery	-
Kuhn et al. <sup>44</sup>	2012	28, F	PML	22	Transseptal <sup>a</sup>	Cyst removal
Paluszkiwicz et al. <sup>45</sup>	2013	80, F	APM	25	NA	Cyst removal
Sun et al. <sup>46</sup>	2013	25, M	AML	NA	No surgery	-
Bonenfant et al. <sup>47</sup>	2014	25, F	AML	19	NA	Cyst removal
Dubey et al. <sup>48</sup>	2017	22, F	PML	NA	NA	Cyst removal, AVR, MVR
Bezak et al. <sup>49</sup>	2019	63, M	AML	24	LA incision	Cyst removal, MVR
Xiao et al. <sup>50</sup>	2019	32, M	APM	46	NA	Cyst removal, MVR
Romano et al. <sup>52</sup>	2009	71, M	PPM	21	NA	Cyst removal, MVr

Abbreviations: AML, anterior mitral leaflet; APM, anterior papillary muscle; AVR, aortic valve replacement; F, female; M, male; MVr, mitral valve repair; MVR, mitral valve replacement; NA, not available; PML, posterior mitral leaflet; PPM, posterior papillary muscle; PTCA, percutaneous transluminal coronary angioplasty.

<sup>a</sup>This patient had a second re sternotomy due to a previous repair of corrected transposition of the great arteries, ventricular septal defect and pulmonary stenosis with a Rastelli-type of procedure followed by pulmonary homograft implantation.

## 3.2 | BCs of the TV

There are nine articles on BCs of the TV, six case reports,<sup>53-59</sup> two images in cardiovascular disease,<sup>57,60</sup> and one letter to the editor,<sup>61</sup> published from 1991 to 2021 (Table 3).

### 3.2.1 | Patient characteristics

Of the nine patients, five were females and four males with an age ranging from 35 to 88 years (mean, 67 ± 15 years). Symptoms at presentation were generally not specific except for few cases with moderate-severe tricuspid regurgitation, with congestive heart failure or symptoms related to other concomitant

cardiac pathology. One patient had an associated muscular ventricular septal defect without hemodynamic significance.<sup>55</sup>

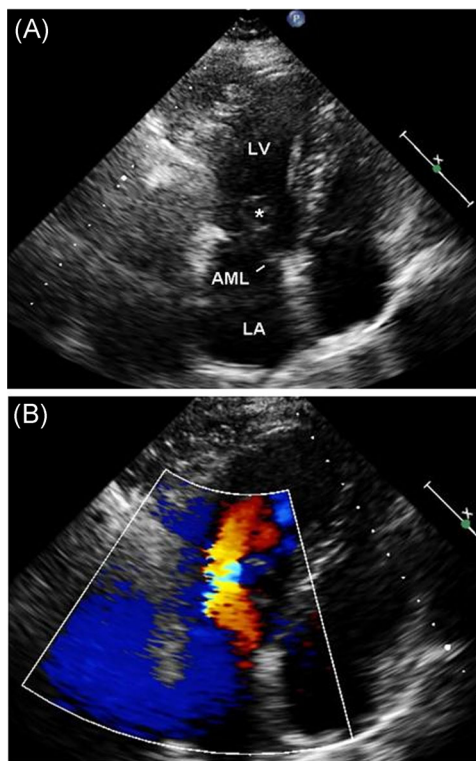
### 3.2.2 | Surgical data

Cyst removal was performed in six cases (55%) while in the remaining three, surgery was considered not indicated. All patients were operated through a median sternotomy using a right atriotomy for cusp excision; in one case even a combined left atrial-transseptal approach is reported. In four patients, BCs removal was associated with various techniques of TV repair; in one patient myocardial revascularization was also performed.

### 3.3 | BCs of the PV

#### 3.3.1 | Patient characteristics

Our search yielded three papers on BCs of the PV, two case reports,<sup>62,63</sup> and one image in cardiovascular disease<sup>64</sup> (Table 4). All



**FIGURE 1** (A) Transthoracic 2D echocardiographic four-chamber view, in a 61-year-old asymptomatic man showing the intracardiac mass (asterisk) attached to the anterior mitral leaflet (AML) and (B) moderate mitral regurgitation. LA, left atrium; LV, left ventricle

were females, 31, 43, and 44 years of age. Auscultation of a cardiac murmur, associated to fatigue and dyspnea, prompted further evaluation with angiographic or echocardiographic disclosure of the BC. In two cases, the pulmonary trunk had an aneurysmal dilatation<sup>62,64</sup> and in one the PV was severely dysplastic.<sup>64</sup>

#### 3.3.2 | Surgical data

Cyst removal was performed in all cases, by a transpulmonary approach. In one patient, PV valvuloplasty was required associated to reduction of the pulmonary trunk size.<sup>64</sup>

### 3.4 | BCs of the AV

We found only a case of a BC, on an images in cardiovascular medicine paper, found on a bicuspid AV in a 22-year-old male with aortic stenosis. The cyst had a maximum diameter of 21 mm with a broad base of implant; it was excised during AVR.<sup>65</sup>

### 3.5 | BC pathology

BCs of the cardiac valves are usually described as round or oval-shaped masses, either bluish or yellowish in color, at times pedunculated and of variable sizes.

Mean maximum size of MV BCs was  $20 \pm 8$  mm, ranging from 3 to 46 mm; in four cases the cyst was considered as “giant” despite a large variability of sizes.<sup>7,9,26,32</sup> The most frequent location of the cysts was the anterior mitral valve leaflet (AML) occurring in 33 cases, followed by the anterior papillary muscle in 8, the posterior mitral leaflet (PML) in 5, and the posterior papillary muscle in 4. The cyst was described to be attached to the chordae tendinae in one, to the mitral annulus in one, and to an unspecified papillary muscle in one; in

**TABLE 3** Characteristics of reported cases of blood cysts of the tricuspid valve

Author, ref. no.	Year	Age, sex	Location	Max. dimension, mm	Surgical approach	Operation
Paşaoğlu et al. <sup>53</sup>	1991	35, M	STL	30	RV	Cyst removal
Timperley et al. <sup>60</sup>	2004	80, F	STL	NA	RA <sup>a</sup>	Cysts removal
Michelena et al. <sup>54</sup>	2007	55, M	ATL	30	RA	Cysts removal, TVr
Agac et al. <sup>55</sup>	2009	72, F	STL	20	No surgery	-
Grapsa et al. <sup>56</sup>	2011	88, F	STL	25	No surgery	-
Butler et al. <sup>57</sup>	2015	66, M	NA	NA	RA	Cysts removal, TVr
Kalçık et al. <sup>58</sup>	2015	65, M	PTL	18	LA-Transseptal	Cysts removal, TVr, CABG
Aydin et al. <sup>59</sup>	2019	68, F	STL	23	No surgery	-
Taylan et al. <sup>61</sup>	2021	74, F	ATL	NA	RA	Cyst removal, TVr

Abbreviations: ATL, anterior tricuspid leaflet; CABG, coronary artery bypass grafting; F, female; M, male; NA, not available; PTL, posterior tricuspid leaflet; RA, right atriotomy; RV, right ventriculotomy; STL, septal tricuspid leaflet; TVr, tricuspid valve repair.

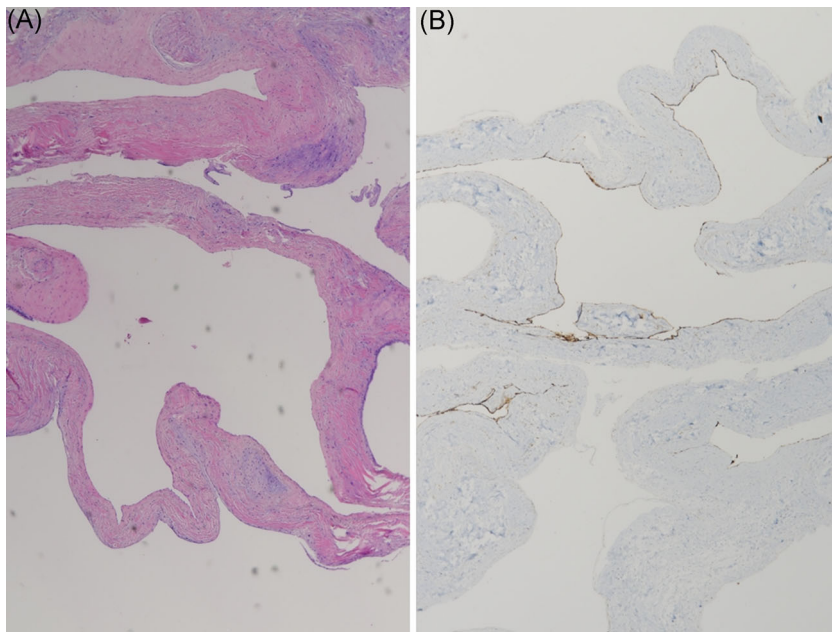
<sup>a</sup>Unless specifically indicated, cyst removal was considered performed through a right atriotomy.

**TABLE 4** Characteristics of reported cases of blood cysts of the pulmonary valve

Author, ref. no.	Year	Age, sex	Location	Max. dimension, mm	Surgical approach	Operation
Liese et al. <sup>62</sup>	1963	31, F	PPC	40	Transpulmonary	Cyst removal
Minato et al. <sup>63</sup>	1997	43, F	RPC	8	Transpulmonary	Cyst removal
Zhang et al. <sup>64</sup>	2021	44, F	NA	30	Transpulmonary	Cyst removal, PV repair, reduction PA plasty <sup>a</sup>

Abbreviations: F, female; PA, pulmonary artery; PPC, posterior pulmonary cusp; PV, pulmonary valve; RPC, right pulmonary cusp.

<sup>a</sup>This patient had a dysplastic pulmonary valve.



**FIGURE 2** (A) Histologic section of the blood cyst wall showing the presence of fibrous tissue (hematoxylin-eosin stain, original magnification  $\times 4$ ). (B) Immunostaining for the endothelial antigen CD31 (brown color) highlighting the endothelial layer on the luminal surface (immunoperoxidase, hematoxylin counterstaining, original magnification  $\times 4$ )

one patient with two BCs both AML and PML were involved.<sup>14</sup> In three patients, two BC were found.<sup>6,14,30</sup> Some BCs have been described as multilobulated.<sup>9,12,17,50</sup>

The maximum size of TV BCs ranged from 18 to 33 mm (mean,  $24 \pm 5$  mm); in three cases the size of the BC was not indicated.<sup>57,60,61</sup> BCs were located on the septal TV leaflet in four, anterior TV leaflet in two, and the posterior TV leaflet in one case; in one patient location of the BC was not reported.<sup>57</sup>

The maximum size of the three PV BCs was 8, 30, and 40 mm, respectively; according to the authors' description BCs were located on the posterior PV cusp in one case and on the right PV cusp in one; in one case the location is not clearly indicated.<sup>64</sup>

Histology of the excised tissue has not been reported in all surgical cases. When available histologic data, regardless of BC location, have been substantially uniform in describing the BC wall as composed mainly by fibrous tissue of various thickness (Figure 2A), at times with a myxoid stroma. In many cases, the inner BC wall was lined with typical endothelium while smooth calcific spots were occasionally seen.<sup>19,49</sup> The fibrous nature of the BC wall is also confirmed by immunohistochemistry (Figure 2B). Interestingly, no ultrastructural images have been reported in BCs excised from cardiac valves of human beings.

## 4 | DISCUSSION

The first description of an intracardiac BC was reported by Elsässer as late as 1844,<sup>66</sup> while Houser and colleagues are credited with the first use of echocardiography to identify such peculiar lesions.<sup>4</sup> In 1968, the first surgical removal of an MV BC was reported by Leatherman et al.<sup>3</sup>; as described by the authors "The cyst was attached to the septal leaflet at the point where the leaflet was joined by chordae tendineae. During excision the cyst was opened and blood drained out. Extracorporeal circulation lasted 18 min, and recovery was uneventful."<sup>3</sup>

Intracardiac BCs are usually incidental necropsy findings or are diagnosed clinically predominantly in children, being extremely rare after the first year of life and particularly in adults.<sup>1,2</sup> They are considered benign tumors, nevertheless, while benign from the histologic point of view, they have been associated to relevant complications such as LVOT obstruction and coronary or systemic embolization with consequent severe sequelae such as stroke or myocardial infarction.<sup>4,5,9,11,15,18,20,48</sup> Transthoracic or transesophageal 2D echo are usually considered adequate to detect intracardiac masses even of small dimensions and the presence of an echo-free space within the mass may usually rise the suspicion of the presence of a BC;



others have considered formation of microbubbles inside the cyst, when using contrast real-time echocardiography, as pathognomonic of the presence of a BC.<sup>12,17</sup> However, sometimes solid cardiac tumors, such as myxomas, may present with similar echocardiographic features, therefore, being difficult to be differentiated from a benign BC.<sup>7,8</sup> Furthermore, it must be underlined that other intracardiac masses must be ruled out such as fibroelastomas or infectious vegetations, which also frequently involve the cardiac valves and, therefore, the diagnosis of a BC is not always straightforward and needs histology to be confirmed. The potential role of magnetic resonance imaging has also been stressed in the diagnostic assessment of such patients particularly because of its specificity in ruling out signs of myocardial infiltration and, therefore, to exclude the presence of a malignant lesion.<sup>26</sup>

From this review, it appears that management of patients with a BC is still controversial.<sup>21</sup> Many advocate surgical excision while according to others serial clinical and echocardiographic surveillance can be adequate especially in the presence of masses of small size to monitor the rate and degree of growth.<sup>15</sup> Indeed, in 30% of the patients analyzed surgery was not considered either for BC small size, clinical stability, patient refusal, or advanced age. However, due to the potential for embolization of even small cardiac masses, including BCs, we believe that surgical removal should always be indicated not only to prevent the risk of complications but also to determine or confirm the exact diagnosis.

The origin of BCs of the cardiac valves is still unknown. The first hypotheses were formulated in the early 1900 by Bayne-Jones, studying the blood vessels of the heart valves.<sup>67</sup> Large studies on hearts of dogs and mainly cows and calves obtained from slaughter houses, have demonstrated the presence of valvular BCs in almost 20% of hearts; the results of histological and ultrastructural evaluations support the hypothesis that BCs most likely derive from dilatation of the thin-walled valvular arteries due to the mechanical stresses induced by the pressure gradient when the atrio-ventricular valves are closed with consequent cyst formation.<sup>68,69</sup> This theory, however, does not explain the occurrence of BCs in low-pressure structures such as the PV.<sup>54</sup> Furthermore, it is not clear whether what observed in animal studies can be also be completely applied to human beings. According to Tsutsui et al., BCs could derive by blood trapped in valvular crevices or microscopic invaginations during development<sup>13</sup>; this might explain the finding of BCs in children but not BCs in adults with previously normal echocardiograms. Other consider BCs to derive from valvular hematomas,<sup>24</sup> or being secondary to endocardial inflammation; accordingly, the few cases observed following previous open heart surgery might indicate an additional risk factor of cardiac surgery in the development of BCs, although this most likely is just an occasional association.<sup>22</sup>

This review has some limitations. Since we have excluded from our analysis cases possibly contained in specific textbooks or pathological reviews and those with an uncertain diagnosis, the number of MV BCs may be underestimated. Even if the American Academy of Pediatrics has identified the upper age limit as 21 years for pediatric patients,<sup>70</sup> we arbitrarily considered 18 years as the lowest age for

this review; elevating this limit would have further reduced the number of recognizable cases. Although some of the data which could be obtained from single cases were not complete, this review provides enough evidence to assess the clinical presentation, diagnostic modalities, and management of patients with a cardiac valve BC found in adult patients.

In conclusion, BCs are rarely found in patients  $\geq 18$  years of age and predominantly affect the MV. Although in most cases they represent an incidental finding, sometimes they are heralded by serious acute complications such as syncope and stroke. For such reason, surgical removal, which is a low-risk procedure and can be performed with minimally invasive techniques, should be advisable for BCs of the MV, even if of small size; on the other hand, BCs in the right heart may be treated conservatively unless they cause TV regurgitation or obstruction to blood flow. The origin of valvular BCs is still quite uncertain but since their histological benign nature is well known further studies on pathogenesis would have only a speculative interest. It is likely that the current widespread use of multimodality imaging will increase the number of intracardiac BC detected and hopefully the present review will help to improve the management of such patients.

#### CONFLICT OF INTERESTS

The authors declare that there are no conflict of interests.

#### ORCID

Uberto Bortolotti  <http://orcid.org/0000-0002-9184-6428>

#### REFERENCES

1. Tyebally S, Chen D, Bhattacharyya S, et al. Cardiac tumors. JACC CardioOncology state-of-the-art review. *J Am Coll Cardiol CardioOnc*. 2020;2:293-311.
2. Zimmerman KG, Paplanus SH, Dong S, Nagle RB. Congenital blood cysts of the heart valves. *Hum Pathol*. 1983;14:699-703.
3. Leatherman L, Leachman RD, Hallman GL, Cooley DA. Cyst of the mitral valve. *Am J Cardiol*. 1968;21:420-430.
4. Hauser AM, Rathod K, McGill J, Rosenberg BF, Gordon S, Timmis GC. Blood cyst of the papillary muscle. Clinical, echocardiographic and anatomic observations. *Am J Cardiol*. 1983;61:612-613.
5. Arnold IR, Hubner PJB, Firmin RK. Blood filled cyst of the papillary muscle of the mitral valve producing severe left ventricular outflow tract obstruction. *Br Heart J*. 1990;63:132-133.
6. Xie SW, Lu CL, Picard MH. Blood cyst of the mitral valve: detection by transthoracic and transesophageal echocardiography. *J Am Ass Echocardiogr*. 1992;5:547-550.
7. Ohmoto Y, Tsuchihashi K, Tanaka S, Shimamoto K, Iimura O. Giant endocardial blood cyst in left ventricle resected by transaortic valve approach. *Chest*. 1993;103:965-966.
8. Pelikan HM, Tsang TS, Seward JB. Giant blood cyst of the mitral valve. *J Am Soc Echocardiogr*. 1999;12:1005-1007.
9. Sharma S, Strauss R, Swanson JS. Giant blood cyst of the mitral valve. *Asian Cardiovasc Thorac Ann*. 2000;8:278-279.
10. Kuvin J, Saha P, Rastegar H, Salomon RN, Pandian N, DeNofrio D. Blood cyst of the mitral valve apparatus in a woman with a history of orthotopic liver transplantation. *J Am Soc Echocardiogr*. 2004;17:480-482.
11. Minneci C, Casolo G, Popoff G, Sulla A, Comin CE, Pedemonti E. A rare case of left ventricular outflow obstruction. *Eur J Echocardiogr*. 2004;5:72-75.

12. López-Pardo F, López-Haldón J, Granado-Sánchez C, Rodríguez-Puras MJ, Martínez-Martínez A. A heart inside the heart: blood cyst of mitral valve. *Echocardiography*. 2008;25:928-930.
13. Tsutsui JM, Pommerantzeff P, Pinto IM, Rochitte CE, Mathias W Jr. Characterization of blood-filled cyst by contrast echocardiography and computed tomography. *J Am Soc Echocardiogr*. 2008;21:777e1-3.
14. Denker M, Jexmark T, Hansen F, Tydén P, Roijer A, Lührs C. Bileaflet blood cysts on the mitral valve in an adult. *Am J Soc Echocardiogr*. 2009;22:1085e5-1088e5.
15. Lodha A, Patel J, Haran M, Sadiq A, Shani J. Blood cyst of the mitral valve: a rare cause of stroke. *Echocardiography*. 2009;26:736-738.
16. Park JP, Song JM, Lee JW, et al. A blood-filled cystic mass attached to the anterolateral papillary muscle. *J Cardiovasc Ultrasound*. 2009;17:16-18.
17. Migliore F, Scarabeo V, Corfini A, Piovesana P. Microbubbles contrast inside the cyst: a pathognomonic sign of mitral valve blood cyst. *J Cardiovasc Med*. 2010;11:294-296.
18. Khan T, El-Sharkawi S, Schafer F. A heart within the heart—a rare congenital cause of an embolic stroke. *BMJ Case Rep*. 2010;2012. <https://doi.org/10.1136/bcr.2011.5027>
19. Park MH, Jung SY, Youn HJ, Jin JY, Lee JH, Jung HO. Blood cyst of subvalvular apparatus of the mitral valve in an adult. *J Cardiovasc Ultrasound*. 2012;20:146-149.
20. Bhatt H, Trinh M, Fisher GW. Mitral valve mass detected on pre-operative transesophageal echocardiogram. *J Cardiothorac Vasc Anesth*. 2013;27:1070-1072.
21. Donndorf P, Bermaoui B, Westohal B, Steinhoff G. Asymptomatic blood cyst of the papillary muscle in an adult undergoing coronary bypass surgery. *Interact Cardiovasc Thorac Surg*. 2013;16:402-404.
22. Ansari E, Akhyari P, Lichtenberg A. Blood cyst of the anterior leaflet of the mitral valve in an asymptomatic adult: is surgery an objective? *Heart Surg Forum*. 2015;18:e196-e197.
23. Halim J, van Schaagen FRN, Rizebos RK, Lalezari S. Giant intracardiac blood cyst: assessing the relationship between its formation and a previous cardiac surgery. *Neth Heart J*. 2015;23:392-394.
24. Madhavan S, Jayaprakash K, Jayaprasad N, Sathish G, George R. Blood cyst of the anterior mitral leaflet causing severe mitral regurgitation. *Baylor Univ Med Center Proc*. 2015;28:363-364.
25. Özmen G, Demir M, Ari H, Aktaş I. A rare association: concomitant presence of mitral valve blood cyst with atrial septal aneurysm and cor triatriatum dexter. *Anatol J Cardiol*. 2015;15:502-503.
26. Yilmaz S, Gül M, Canpolat U, Gölhasi Z. Blood cyst of the mitral valve in an adult. *Herz*. 2015;40:329-331.
27. Ahmad F, Barbant S, Levy T. Giant blood cyst of the mitral valve in a disseminated tuberculosis patient. *Echocardiography*. 2016;33:920-922.
28. Okamoto K, Kudo M, Hayashi K, Shimizu H. Endoscopic-assisted minimally invasive resection of a papillary muscle blood cyst in an adult patient. *Interact Cardiovasc Thorac Surg*. 2016;22:231-232.
29. Akutsu H, Muraoka A, Misawa Y. Left ventricular blood cyst in an adult case. *Clin Case Rep*. 2017;5:1441-1443.
30. Pavsic N, Dolenc-Strazar Z, Cercek AC, Klokocovnik T, Prokselj K. Coronary artery embolism from a blood cyst of the mitral valve. *Heart, Lung Circ*. 2017;26:e118-e120.
31. Bagheri A, Ansariaval Z, Khani M, Shekerkhar S, Baghayi R, Sharifi T. Two adherent blood cysts in subvalvular apparatus of the mitral valve causing severe left ventricular outflow tract obstruction. *J Cardiol Cases*. 2018;18:78-80.
32. Ludhwani D, Sheikh B, Sheikh Y. Evaluation of mitral apparatus blood cyst: a case report and review of the literature. *Cureus*. 2019;11:e5612. <https://doi.org/10.7759/cureus.5812>
33. Ma X, Zeng X, Wu Y, et al. Mitral valve blood cyst occlusion of the aortic valve leading to syncope: detection by transthoracic and contrast echocardiography. *J Int Med Res*. 2019;47:3487-3490.
34. Cerik IB, Bektas O, Gunaydin Y, Dereli S, Kaya A. Is surgery really necessary in blood cysts? *Middle Black Sea J Health Sci*. 2020;6:272-274.
35. Ramirez-Mesias DC, Contreras-Valero JF, Pinilla-Monsalve GD, Vesga-Reyes CE. Blood cyst of the mitral valve diagnosed in an adult after systemic thrombolysis. *Case Rep Cardiol*. 2020;2020:4320269. <https://doi.org/10.1155/2020/4320269>
36. Beale RA, Russo R, Beale C, et al. Mitral valve blood cyst diagnosed with the use of multimodality imaging. *Cardiovasc Imag Case Rep*. 2021;5:173-176.
37. Ionac I, Mornos C, Feier H, Derma A, Ionac A. Blood cyst of the mitral valve—is it common? Case report. Romanian. *J Cardiol*. 2021;31:111-115.
38. Zhang X, Zhang L, Li L, Ren T, Jiang S. Totally endoscopic surgical resection for a blood cyst originated from the mitral valve: a case report. *J Cardiothorac Surg*. 2021;16:164. <https://doi.org/10.1186/s13019-021-01535-6>
39. Kurtoğlu N, Yildirim T, Uzunlar B, Çevik C, Incedere O, Dindar I. A rare case: blood cyst of the mitral valve. *Anadolu Kardiyol Derg*. 2005;5:346.
40. Yamamoto H, Nakatani S, Niwaya K, Ohnishi T, Uematsu M, Kitakaze M. Giant blood cyst of the mitral valve: echocardiographic and intraoperative images. *Circulation*. 2005;112:e341.
41. Akinci S, Celikyurt U. Mitral valve blood cyst with regurgitation. *Heart Asia*. 2010;2:112. <https://doi.org/10.1136/ha.2010.002162>
42. Combaret N, Azarnoush K, Marcaggi X, et al. A blood cyst on the anterior mitral leaflet complicated by cerebrovascular stroke. *J Heart Valve Dis*. 2012;21:688.
43. Grimaldi A, Capritti E, Pappalardo F, et al. Blood cyst of the mitral valve. *J Cardiovasc Med*. 2012;13:46.
44. Kühn A, Schreiber C, Weirich G, Vogt M. Intracardiac blood cyst: rare finding in a complex congenital heart lesion. *Eur Heart J Cardiovasc Imag*. 2012;33:229.
45. Paluszkiwicz L, Morshuis M, Koster A, Antończyk K, Gummert J. A large blood cyst of the mitral valve. *Eur Heart J Cardiovasc Imag*. 2013;14:315.
46. Sun F, Ren W, Bi W, Zhang Y, Guo BA. "Balloon" on the mitral valve. *J Am Coll Cardiol*. 2013;62:81.
47. Bonenfant F, Essadiqi B, Gahide G, Jeanmart H, Farand P. Mitral valve blood cyst detected by multimodality imaging. *Can J Cardiol*. 2014;30:1109.e5-1109.e6.
48. Dubey G, Shukla A, Madan T. Rheumatic heart disease with a mitral blood cyst: three-dimensional echocardiographic imaging. *Heart Asia*. 2017;9:1.
49. Bezak B, Artemiou P, Hudec V, Hulman M. Blood cyst of the anterior mitral leaflet causing severe mitral regurgitation. *J Card Surg*. 2019;34:1354-1355.
50. Xiao W, He Y, Xu M, Hua Z, Song L. A giant blood cyst of mitral valve resulting in severe left ventricular outflow tract obstruction. *Ann Thorac Surg*. 2019;107:e139.
51. Wang J, Han J, Zhang Y, et al. Diagnosis of cardiac blood cyst by echocardiography in 8 cases. *Authorea* 2020; <https://doi.org/10.22541/au.159654485.57477936>
52. Romano S, Krishnasamy K, Jariwala N, Chung J, Farzaneh-Far A. Giant mitral blood cyst: cardiovascular magnetic resonance imaging features. *Int J Cardiol*. 2016;203:17-18.
53. Paşaoğlu I, Doğan R, Nazlı N, Güngen Y, Bozer AY. Blood cyst originating from tricuspid septal leaflet. *J Cardiovasc Surg*. 1991;32:589-591.
54. Michelena HI, Mulvagh SI, Schaff HV, Enriquez-Sarano ML, Klarich KW. A heart-shaped mass inside a heart: echocardiographic diagnosis, pathology, and surgical repair of a flail tricuspid valve caused by a large blood-filled cyst. *J Am Soc Echocard*. 2007;20:771.e3-6.



55. Agac MT, Acar Z, Turan T, Karadag B, Kul S, Erkan H. Blood cyst of a tricuspid valve: an incidental finding in a patient with ventricular septal defect. *Eur J Echocardiogr*. 2009;10:588-589.
56. Grapsa J, Cheung WS, Dawson D, Nihoyannopoulos P. Congenital tricuspid valve blood cyst: imaging with three-dimensional contrast echocardiography. *Hellenic J Cardiol*. 2011;52:278-280.
57. Butler TC, Wall D, Hamilton-Craig C. A ring in the heart. *Heart*. 2015; 101:1965.
58. Kalçık M, Yesin M, Gürsoy MO, et al. An unusual mass of tricuspid valve in an adult patient: blood-filled cyst. *Echocardiography*. 2015; 32:1199-1202. <https://doi.org/10.1111/echo.12878>
59. Aydin C, Engin M, Kul S. Tricuspid regurgitation due to blood cyst. *Echocardiography*. 2019;36:2108-2109.
60. Timperley J, Forfar C, Becher H, Pillai R. Heart within a heart. Contrast 3-dimensional echocardiography imaging of a tricuspid valve blood cyst. *Circulation*. 2004;110:e324-e325.
61. Taylan G, Ustabaşoğlu FE, Binboğa B, Manav A, Yüksel V. An unusual cardiac mass associated with severe tricuspid valve regurgitation. *Balkan Med J*. 2021;38:195-197.
62. Liese GJ, Brainard SC, Goto U. Giant blood cyst of the pulmonary valve. *New Engl J Med*. 1963;269:465-467.
63. Minato H, Manabe T, Masaki H, Kawahara Y. Blood cyst of the pulmonary valve in an adult: report of a case and review of the literature. *Human Pathol*. 1997;28:252-255.
64. Zhang W, Yan D, Xie C, et al. Giant blood cyst, isolated dysgenesis of pulmonary valve and pulmonary artery aneurysm: a rare combination. *Int J Cardiovasc Imag*. Published online July 25, 2021. <https://doi.org/10.1007/s10554-021-02288-y>
65. Gu J, Chen Y, Zhang H, Meng W, Hu J, Zhang E. Multimodality images of a giant blood cyst originating from the bicuspid aortic valve. *Circulation*. 2014;130:e165-e166.
66. Elsässer C. Bericht über di Ereignisse in der Gebäranstalt des Catherinen Hospitals im Jahre 1844. *Med Correspondenzblatt*. 1844;14: 297.
67. Bayne-Jones S. The blood-vessels of the heart valves. *Am J Anat*. 1917;21:449-463.
68. Takeda T, Makita T, Nakamura N, Kimizuka G. Morphologic aspects and morphogenesis of blood cysts on canine cardiac valves. *Vet Pathol*. 1991;28:16-21.
69. Marcato PS, Benazzi C, Bettini G, et al. Blood and serous cysts in the atrioventricular valves of the bovine heart. *Vet Pathol*. 1996;33:14-21.
70. Peykoff Hardin A, Hackell JM. Age limits of pediatrics. *Pediatrics*. 2017;140:e20172151.

**How to cite this article:** Bortolotti U, Vendramin I, Lechiancole A, et al. Blood cysts of the cardiac valves in adults: Review and analysis of published cases. *J Card Surg*. 2021;1-9. <https://doi.org/10.1111/jocs.15992>