Case Report

Hazem El Beyrouti, Martin Oberhoffer, Angela Kornberger, Andres Beiras-Fernandez* and Christian-Friedrich Vahl

Acute heart failure due to giant left atrium: remote ECLS implantation for interhospital transfer and bridging to decision

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Abstract

Background: Remote extracorporeal life support (ECLS) implantation and mobile ECLS are increasingly applied to enable the treatment of patients with refractory heart failure at specialized care centers. Giant left atrium (GLA) is a rare cause but may lead to acute and devastating heart failure.

Case report: Acute heart failure that gave rise to a prolonged period of out-of-hospital cardiopulmonary resuscitation in a patient who underwent remote ECLS implantation was found to have resulted from a GLA of impressive dimensions. There having no reasonable option for immediate surgical therapy, the patient was successfully bridged to recovery with a view to subsequent heart transplantation.

Conclusions: The threshold to rapid institution of ECLS should be low in patients with acute refractory heart failure, including those with uncommon pathologies. Remote ECLS implantation and interhospital transfer on mobile ECLS increase the chances of survival in cases requiring treatment by specialized care centers.

Keywords: extracorporeal life support; giant left atrium; heart failure; mobile ECLS; remote ECLS implantation.

Introduction

Giant left atrium (GLA) is commonly defined as a left atrium with a diameter of \geq 65 mm and found most

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frequently as a result of mitral valve disease. The condition may be asymptomatic or manifest itself by different symptoms, including atrial fibrillation, thrombus formation, and heart failure [1, 2].

Surgical therapy aims at reducing the size of GLA and relies on a variety of techniques for plicating or resecting parts of the left atrium [1]. Due to the difficulty of exposing a heart with a left atrium of excessive dimensions, the surgical therapy of GLA was variously reported to have required (partial) autotransplantation [3, 4].

With regard to extracorporeal life support (ECLS), there is wide agreement not only on its use in patients with severe heart failure refractory to conventional therapy but also that it should be instituted before the onset of multiorgan failure [5]. In patients with acute refractory heart failure or after cardiopulmonary resuscitation (CPR) in whom diagnosis and/or neurological status are not clear or the further course of therapy is yet to be determined, ECLS may serve as a bridge to decision. Over the past decade, remote ECLS implantation and interhospital transfer on mobile ECLS have come to play an increasing role [6].

Case report

A 57-year-old male with acute heart failure was admitted to a community hospital after return of spontaneous circulation (ROSC) following cardiac arrest and approximately 65 min of out-of-hospital CPR. Although his cardiac rhythm at the time of ROSC was not documented, he was in atrial fibrillation upon arrival at the community hospital. It turned out that he had undergone redo mitral valve replacement with a mechanical valve and tricuspid valve repair 5 years preceding the event. Nearly two decades earlier, he had undergone mitral valve replacement for mitral regurgitation in the setting of cardiomegaly. Echocardiography (ECG) and computed tomography

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(CT) showed a grotesque case of GLA (Figures 1 and 2) that had gone undiagnosed because the patient had not presented for follow-up examinations after the most recent surgical procedure (i.e. for 5 years). ECG findings included sludge in GLA and a left ventricular ejection fraction of 10%. Fluoroscopy showed a normal motion of mitral valve leaflets.

When the patient remained in severe cardiogenic shock, our mobile ECLS team was called for ECLS implantation and subsequent air transport to enable further



Figure 1: Transversal section of GLA on initial CT after cardiac arrest and CPR (dimensions: 240×176 mm).



Figure 2: Frontal section of GLA on initial CT.

treatment at our tertiary care center. The inflow and outflow for the ECLS system (Stöckert Centrifugal Pump and Centrifugal Pump Console) were instituted by percutaneous placement of a 17 Fr NovaPort cannula in the left femoral artery and a 21 Fr Biomedicus cannula in the left femoral vein.

Both ECLS implantation and interhospital transfer were implemented without complications. In the further course, preventing thromboembolic complications while at the same time controlling an increasingly dramatic hemorrhagic tendency turned out to be a challenge. ECG showed spontaneous contrast/sludge in the left atrium (Figure 3), although the patient's initial international normalized ratio (INR) of 3.3 was accompanied by moderate thrombocytopenia and his partial thromboplastin time (PTT) was 50.2 s after receiving 2000 IU unfractionated heparin for ECLS implantation. Over the next 2 days, his INR climbed to 8.9 and extensive spontaneous oropharyngeal, pulmonary, and gastrointestinal bleeding required transfusion of several units of packed red blood cells and moderate correction of his coagulation status.

Starting at 2800 rpm, the ECLS system provided sufficient blood flow (of \sim 4 L) to keep up adequate perfusion. Sedation was terminated and the patient was weaned from catecholamines within 48 h while his renal and hepatic functions simultaneously recovered. He woke up and turned out to be responsive though slightly delirious.

Surgical options were evaluated but appeared little promising due to the very size of GLA and the patient's risk constellation. Therefore, it was resolved to wean him from ECLS and retransfer him to the community hospital for further recovery and subsequent evaluation for heart transplantation. The ECLS system was kept at about 2800 rpm for about 24 h and at 1900–2200 for another

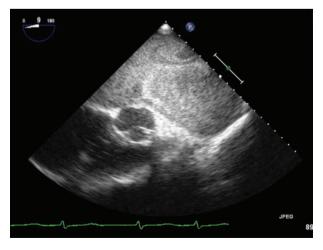


Figure 3: ECG view of GLA with spontaneous contrast/sludge.

period of about 24 h. Then, the pump speed was reduced to 1500 rpm (1.3 L) before the ECLS system was finally removed a few hours later.

Early mobilization was initiated and mechanical respiration was terminated as soon as oropharyngeal and pulmonary bleeding were under control. Intravenous (i.v.) heparin was continued to prevent thrombus formation within GLA, and after a total intensive care unit stay of 6 days, the patient was retransferred and arrived in stable condition at the community hospital. At the time of retransfer, he was on i.v. unfractionated heparin (PTT 59.9 s) and had an INR of 1.7.

Follow-up for the present case report revealed that he had taken a fatal course due to mesenteric ischemia suspected to have been caused by thromboembolism from GLA within a few days from arriving at the community hospital.

Discussion

The relevance of heart failure in patients with GLA was demonstrated by Kim et al. [2], who followed a cohort of 163 patients up for a median of 22 months. Eighteen (11%) patients were admitted to hospital due to heart failure, and the fact that nearly 30% of those hospitalized for heart failure in this series took a fatal course is in keeping with the devastating character of the condition we observed in our patient.

Mortality is moreover known to be extremely high in patients who sustained out-of-hospital cardiac arrest. Survival-to-discharge after adult out-of-hospital cardiac arrest in Europe was found to be as low as 9% by a review of 67 prospective studies [7], whereas register data from the United States suggested a survival of 8.3% for the year 2012 [8]. Given the low probability of surviving out-of-hospital cardiac arrest and the severity of our patient's underlying pathology, we conclude that an early institution of ECLS made a relevant contribution to his initial survival and recovery.

Simultaneously, our case illustrates the advantages of remote ECLS implantation and interhospital transfer on mobile ECLS. The benefits of pretransfer institution of and transport on ECLS we saw in our patient are in keeping with an increasing number of reports on interhospital transfer on ECLS, suggesting favorable results and demonstrating that pretransfer ECLS implantation enables an immediate improvement of cardiorespiratory parameters, prevents delays in hemodynamic stabilization, and increases transport safety [9–11]. The fact that our patient's neurological status was unclear and, in combination with his extremely unfavorable cardiac anatomy and high-risk profile, required careful evaluation of surgical therapy options addressing GLA on the one hand and candidacy for heart transplantation on the other illustrates the notion of ECLS as a bridge to decision. The ECLS thus served not only to achieve hemodynamic stability but also to gain time to evaluate neurological recovery, investigate medical issues and therapy options, and obtain information on questions such as social support and patient consent to further therapies.

Conclusion

In summary, the fast recovery our patient made on ECLS leads us to conclude that the threshold to rapid institution of mechanical circulatory support should be low in patients with acute refractory heart failure, including those with unusual and rare pathologies. We conclude that ECLS weaning should be attempted and that surgical therapy options including heart transplantation should be carefully evaluated. Sufficient anticoagulation is mandatory. The fact that follow-up revealed subsequent death from thromboembolic complications highlights the danger of thromboembolism inevitably associated with GLA.

Author Statement

Funding: Authors state no funding involved. Conflict of interest: Authors state no conflict of interest. Informed consent: Informed consent has been obtained from the individual's relatives.

Author Contributions

Andres Beiras-Fernandez: Conceptualization; Formal analysis; Supervision; Validation; Writing – review & editing; Hazem El Beyrouti: Conceptualization; Data curation; Formal analysis; Methodology; Supervision; Martin Oberhoffer: Conceptualization; Data curation; Supervision; Writing – review & editing; Angela Kornberger: Supervision; Validation; Writing – original draft; Christian-Friedrich Vahl: Conceptualization; Validation; Writing – review & editing.

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Supplementary Material: The article (https://doi.org/10.1515/iss-2018-0029) offers reviewer assessments as supplementary material.

Reviewer Assessment

Hazem El Beyrouti, Martin Oberhoffer, Angela Kornberger, Andres Beiras-Fernandez* and Christian-Friedrich Vahl

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Reviewers' Comments to Original Submission

Reviewer 1: anonymous

Sept 25, 2018

Reviewer Recommendation Term:	Accept with Minor Revision
Overall Reviewer Manuscript Rating:	70
Custom Review Questions	Response
Is the subject area appropriate for you?	4
Does the title clearly reflect the paper's content?	4
Does the abstract clearly reflect the paper's content?	4
Do the keywords clearly reflect the paper's content?	5 - High/Yes
Does the introduction present the problem clearly?	4
Are the results/conclusions justified?	3
How comprehensive and up-to-date is the subject matter presented?	3
How adequate is the data presentation?	2
Are units and terminology used correctly?	4
Is the number of cases adequate?	N/A
Are the experimental methods/clinical studies adequate?	N/A
Is the length appropriate in relation to the content?	4
Does the reader get new insights from the article?	3
Please rate the practical significance.	2
Please rate the accuracy of methods.	N/A
Please rate the statistical evaluation and quality control.	N/A
Please rate the appropriateness of the figures and tables.	3
Please rate the appropriateness of the references.	4
Please evaluate the writing style and use of language.	4
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	Yes

Comments to Authors:

The submitted Case report describes an interesting situation of remote ECLS implantation resulting in a fast initial recovery of cardiac function and weaning from ECLS in a patient with giant left atrium.

However, the following facts remain unclear and should be completed: Section "Case report":

- How long was the time of amateur and professional CPR out of hospital?

- What was the initial ECG-rhythm observed on CPR?

- Is there any data of echocardiographic parameters (ejection fraction, valve function) and/or coronary status prior to the cardiac arrest, e.g.

in the setting of follow up examination after re-do mitral-valve repair?

- Is there any data of echocardiographic parameters (ejection fraction, valve function) after ROSC in refractory cardiogenic shock?

- How where the parameters of ECLS therapy (Flow, MAP, Pulsatility)?

- Was there any remaining atrial sludge in echocardiography at the time of referral to the community hospital and how was the INR? Section "Conclusion"

- It is well known that ECLS implantation can be a successful tool for bridge to decision. Due to this rare case of giant left atrium and ECLS implantation: What is your specified conclusion for ECLS weaning and further therapy in this group of patients?

Reviewer 2: anonymous

Nov 05, 2018

Reviewer Recommendation Term:	Accept with Minor Revision
Overall Reviewer Manuscript Rating:	50
Custom Review Questions	Response
Is the subject area appropriate for you?	5 - High/Yes
Does the title clearly reflect the paper's content?	4
Does the abstract clearly reflect the paper's content?	4
Do the keywords clearly reflect the paper's content?	5 - High/Yes
Does the introduction present the problem clearly?	5 - High/Yes
Are the results/conclusions justified?	5 - High/Yes
How comprehensive and up-to-date is the subject matter presented?	4
How adequate is the data presentation?	4
Are units and terminology used correctly?	5 - High/Yes
Is the number of cases adequate?	N/A
Are the experimental methods/clinical studies adequate?	N/A
Is the length appropriate in relation to the content?	4
Does the reader get new insights from the article?	3
Please rate the practical significance.	3
Please rate the accuracy of methods.	4
Please rate the statistical evaluation and quality control.	N/A
Please rate the appropriateness of the figures and tables.	4
Please rate the appropriateness of the references.	3
Please evaluate the writing style and use of language.	4
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	Yes

Comments to Authors:

Interresting, well written case report reflecting the current use of ECLS; however, the pathology is unusual and rare and therefore interresting to read; the standards of ECLS are mentioned and discussed, a more detailed technical information on the ECLS setup and components might add valuable additional information, especially considering the air transportation on ECLS.

Authors' Response to Reviewer Comments

Nov 19, 2018

1. The total duration of CPR was 60 - 65 minutes.

2. The initial rhythm upon ROSC was not documented. Upon arrival at the community hospital, the patient was in atrial fibrillation.

3. The patient had not complied with his follow-up schedule after valve surgery. He had not seen a cardiologist after valve replacement, i.e. for 5 years.

4. The ejection fraction documented at the community hospital was 10%. Fluoroscopy showed normal motion of the mitral valve leaflets.

5. The ECLS system was started at a speed of 2800 RPM (approx. 4 L), and these settings were left unchanged for approx. 24 hours. This was followed for approx. 24 hours at a reduced speed of 1900–2200 RPM, before the pump speed was finally reduced to 1500 rpm (1.3 L) for a few hours before explantation. Pressure upon arrival at our ICU was 95/65 mmHg (MAP 70 mmHg). After a few hours, pulsatility increased, and MAP was kept between 66 and 80 mmHg throughout the entire period on ECLS. This was interrupted by short MAP increases to up to 100 mmHg in the course of catecholamine dose adjustments.

6. By the time when the patient was referred back to the community hospital, the amount of sludge in the left atrium had decreased significantly but echocardiography still a certain measure of smoke in the left atrium. The patient's INR at the time of retransfer was 1.7, but he was on i.v. unfractionated heparin (PTT 59.9 seconds).

7. We conclude that ECLS weaning should be attempted and that surgical therapy options including HTX should be evaluated carefully. Sufficient anticoagulation is mandatory. The ECLS setup consisted of a Stöckert Centrifugal Pump and Centrifugal Pump Console. Inflow and outflow for the ECLS system were instituted by percutaneous placement of a 17 Fr NovaPort cannula in the left femoral artery and a 21 Fr Biomedicus cannula in the left femoral vein.

Reviewers' Comments to Revision

Reviewer 1: anonymous

Nov 24, 2018

Reviewer Recommendation Term:	Accept	
Overall Reviewer Manuscript Rating:	80	
	-	
Custom Review Questions	Response	
Is the subject area appropriate for you?	4	
Does the title clearly reflect the paper's content?	5 - High/Yes	
Does the abstract clearly reflect the paper's content?	4	
Do the keywords clearly reflect the paper's content?	4	
Does the introduction present the problem clearly?	4	
Are the results/conclusions justified?	5 - High/Yes	
How comprehensive and up-to-date is the subject matter presented?	3	
How adequate is the data presentation?	4	
Are units and terminology used correctly?	5 - High/Yes	
Is the number of cases adequate?	N/A	
Are the experimental methods/clinical studies adequate?	4	
Is the length appropriate in relation to the content?	5 - High/Yes	
Does the reader get new insights from the article?	2	
Please rate the practical significance.	2	
Please rate the accuracy of methods.	5 - High/Yes	
Please rate the statistical evaluation and quality control.	N/A	
Please rate the appropriateness of the figures and tables.	4	
Please rate the appropriateness of the references.	4	

Please evaluate the writing style and use of language.	4
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	Yes

Comments to Authors:

No further comments.

Reviewer 2: anonymous

Nov 21, 2018

Reviewer Recommendation Term:	Accept	
Overall Reviewer Manuscript Rating:	45	
Custom Review Questions	Response	
Is the subject area appropriate for you?	5 - High/Yes	
Does the title clearly reflect the paper's content?	5 - High/Yes	
Does the abstract clearly reflect the paper's content?	5 - High/Yes	
Do the keywords clearly reflect the paper's content?	5 - High/Yes	
Does the introduction present the problem clearly?	5 - High/Yes	
Are the results/conclusions justified?	5 - High/Yes	
How comprehensive and up-to-date is the subject matter presented?	5 - High/Yes	
How adequate is the data presentation?	5 - High/Yes	
Are units and terminology used correctly?	5 - High/Yes	
s the number of cases adequate?	N/A	
Are the experimental methods/clinical studies adequate?	N/A	
s the length appropriate in relation to the content?	5 - High/Yes	
Does the reader get new insights from the article?	3	
Please rate the practical significance.	4	
Please rate the accuracy of methods.	5 - High/Yes	
Please rate the statistical evaluation and quality control.	N/A	
Please rate the appropriateness of the figures and tables.	5 - High/Yes	
Please rate the appropriateness of the references.	4	
Please evaluate the writing style and use of language.	5 - High/Yes	
Please judge the overall scientific quality of the manuscript.	3	
Are you willing to review the revision of this manuscript?	Yes	

No further comments.