Clinical Case Reports

Open Access

CASE REPORT

A case of successful bystander cardiopulmonary resuscitation of an adult with Bland-White-Garland syndrome

Hidetada Fukuoka[1], Tetsuya Watanabe[1], Yukinori Shinoda[1], Kuniyasu Ikeoka[1], Tomoko Minamisaka[1], Hirooki Inui[1], Keisuke Ueno[1], Soki Inoue[1], Kentaro Mine[1], Koichi Toda[2], Yoshiki Sawa[2] & Shiro Hoshida[1]

[1]Department of Cardiovascular Medicine, Yao Municipal Hospital, 1-3-1 Ryuge-cho, Yao, Osaka 581-0069, Japan
[2]Department of Cardiovascular Surgery, Osaka University Graduate School of Medicine, 2-2 Yamadaoka, Suita, Osaka 565-0871, Japan

Correspondence
Shiro Hoshida, Department of Cardiovascular Medicine, Yao Municipal Hospital, 1-3-1 Ryuge-cho, Yao, Osaka 581-0069, Japan. Tel: +81-72-922-0881; Fax: +81-72-924-4820; E-mail: shiro.hoshida@hosp-yao.osaka.jp

Funding Information
No sources of funding were declared for this study.

Received: 19 July 2017; Revised: 17 August 2017; Accepted: 25 August 2017

Clinical Case Reports **2017; 5(11): 1797–1801**

doi: 10.1002/ccr3.1183

Key Clinical Message

In Japan and worldwide, the increase in educational interventions about resuscitation training significantly increases favorable neurological survival in out-of-hospital cardiopulmonary arrest cases treated with bystander cardiopulmonary resuscitation (BCPR) each year. This case with Bland-White-Garland syndrome having high-quality BCPR by nurses demonstrates the importance of education about BCPR.

Keywords

Bland-White-Garland syndrome, cardiopulmonary resuscitation.

Introduction

Bland-White-Garland (BWG) syndrome, a rare congenital heart disease in which the left coronary artery (LCA) originates from the pulmonary artery (PA), comprises approximately 0.3% of all congenital cardiac anomalies [1, 2]. Approximately 90% of cases of BWG syndrome occur in infants, many of whom die of myocardial infarction and lethal arrhythmia in early childhood. However, these complications are caused by BWG syndrome in adults who had an asymptomatic infancy [3].

Cardiopulmonary resuscitation by a bystander (BCPR) increases patient survival after out-of-hospital cardiopulmonary arrest (OHCA). In Japan, following the spread of educational intervention about resuscitation training, the number of patients who experience BCPR and survive after OHCA has increased annually [4].

Here, we describe a case of BWG syndrome in a woman who was resuscitated from a complicated lethal arrhythmia by BCPR without subsequent neurological impediments.

Case Presentation

A 33-year-old woman with a history of three vaginal births (8, 9, and 10 years prior), but no other prior medical history had felt chest discomfort at work for 3 years. After a year-end party with colleagues in December 2015, she started to go home on a bicycle accompanied by two female coworkers. On the way, she suddenly lost consciousness. Finding no pulse or signs of breathing, her coworkers immediately called emergency medical services (EMS) and initiated chest compressions and mouth-to-mouth respirations. The EMS arrived 9 min after the cardiopulmonary arrest (CPA) and attached and activated an automated external defibrillator (AED). Defibrillation was successful, resulting in termination of ventricular fibrillation (VF) (Fig. 1). Her circulation spontaneously returned 14 min after CPA. She was subsequently transferred to our hospital.

Upon arrival at the hospital, her consciousness was three points (E1V1M1) on the Glasgow Coma Scale. Her spontaneous breathing was weak, so she was intubated

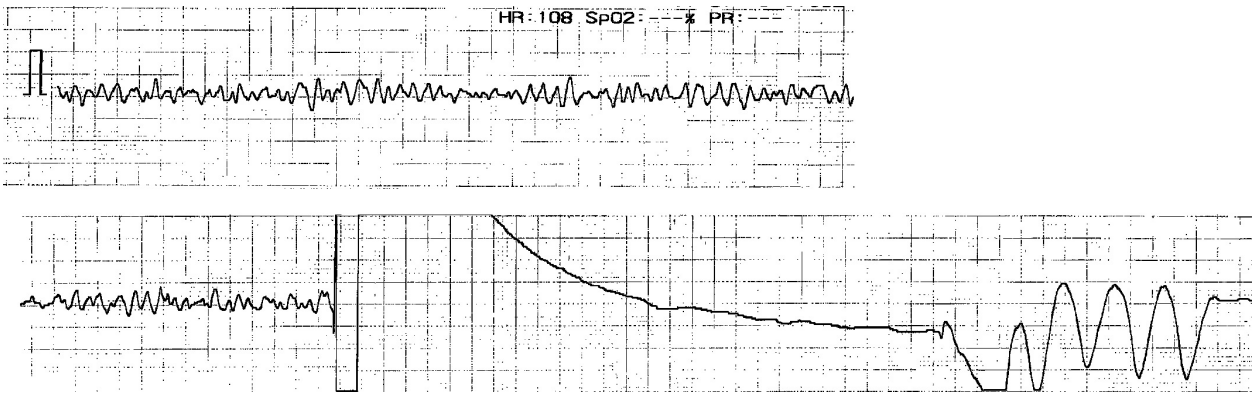

Figure 1. Automated external defibrillator monitor. Ventricular fibrillation and termination by defibrillation of 150 J.

and connected to a respirator. Blood values were within the normal range, except for positive human fatty acid-binding protein. Chest radiography, cerebral computed tomography, and subsequent cerebral magnetic resonance imaging findings were normal. Electrocardiography (ECG), a few hours after admission, showed normal sinus rhythm without Brugada-like electrocardiography changes or QT prolongation (QTc, 0.466s). Transthoracic echocardiography revealed normal left ventricle wall motion with an ejection fraction of 60% and no significant valvular disease. Neither hypertrophic cardiomyopathy nor arrhythmogenic right ventricular dysplasia was suspected.

After admission, she had a good clinical course with no lethal arrhythmic recurrence. We completed her intensive care management on day 5. Fortunately, she had no neurological impediments.

Coronary angiography demonstrated an anomalous arising of the LCA from the PA with retrograde filling from an enlarged and meandering right coronary artery (RCA) through collateral vessels that developed in the interventricular septum without stenosis (Fig. 2). We diagnosed her with BWG syndrome. Technetium-99 m single-photon emission computed tomography (99mTc-SPECT) demonstrated significant hypoperfusion without defects after exercise and incomplete fill-in at rest in the anterior wall and apex (Fig. 3A). During exercise, arrhythmia was absent, but ECG showed ST-segment depression in the II, III, and a VF leads with chest discomfort and cold sweating. We concluded that all symptoms and the lethal arrhythmia were complications of myocardial ischemia by the BWG syndrome. For radical cure, she underwent patch closure of the orifice of the left main tract from the main PA and coronary artery bypass surgery. Postoperatively, 99mTc-SPECT demonstrated improvement in the ischemic areas (Fig. 3B). During exercise, arrhythmia, ST-segment change, and symptoms were absent. Her postoperative course was good and she returned to her normal life.

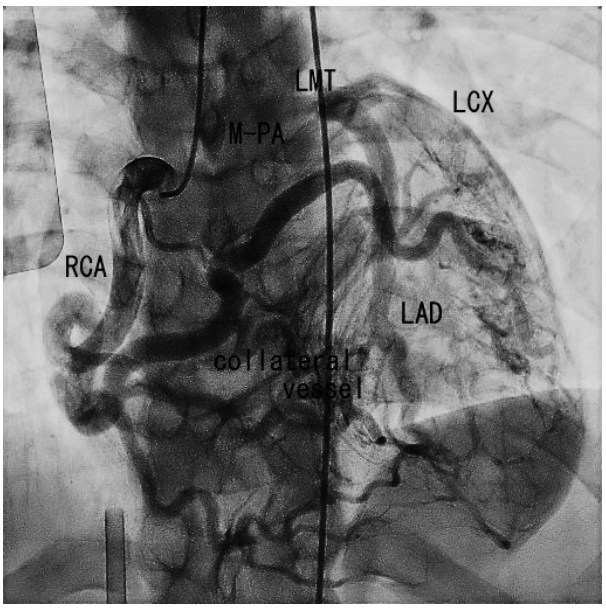

Figure 2. Coronary angiography cranial 30°. Anomalous arising of the left coronary artery from the pulmonary artery with retrograde filling from an enlarged and meandering right coronary artery through the collateral vessel developed in interventricular septum. LCX, left circumflex artery; LMT, left main coronary trunk; M-PA, main pulmonary artery.

Discussion

Bland-White-Garland syndrome is a congenital cardiac anomaly in which the LCA originates from the PA. The abnormality of the anatomical relationship between the aorticopulmonary septum and the coronary opening position during fetal development is the cause of BWG syndrome [5]. It is a rare disease with an assumed incidence of 1 in every 300,000 births [6]. Approximately 90% patients with this syndrome have an ischemic disorder in

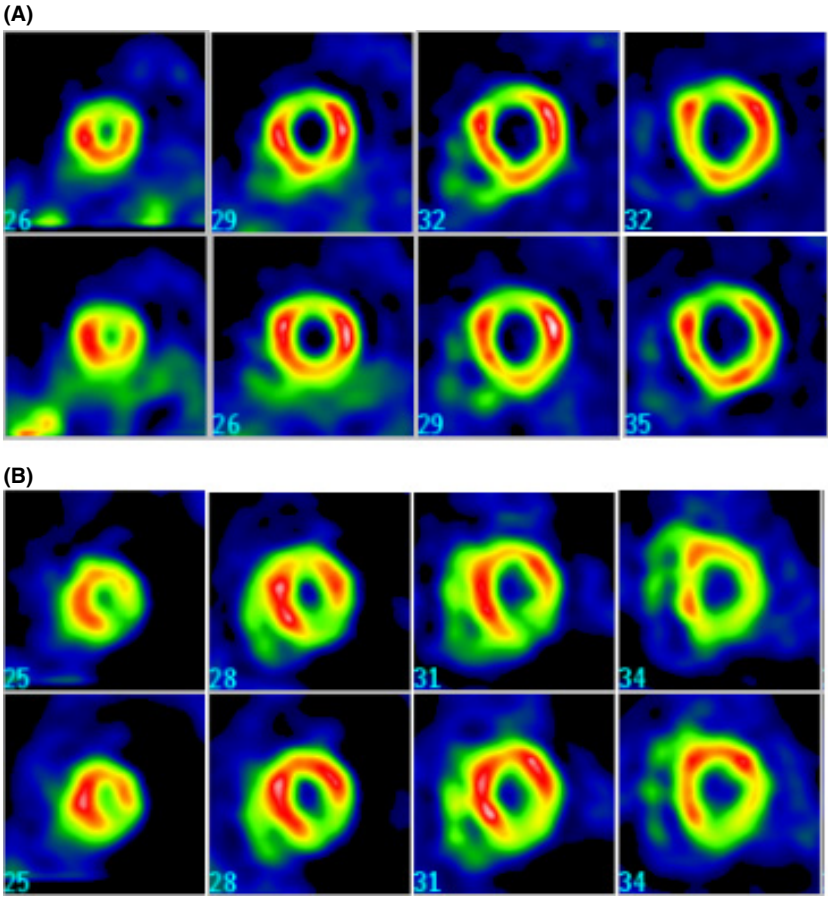

Figure 3. Technetium-99 m single-photon emission computed tomography (upper, stress; lower, rest). (A) Preoperative study showing hypoperfusion without defect and incomplete fill-in at rest in the anterior wall and apex. (B) Postoperative study showing significantly improved perfusion in the anterior wall and apex and a slight decrease in accumulation within the inferoposterior wall without fill-in. We cannot deny the myocardial disorder in the inferoposterior wall, but the quantitative gated single-photon emission computed tomography showed normal wall motion with an ejection fraction of 65%.

the LCA because of insufficient blood flow from the collateral pathways. Complications such as ischemic heart disease, lethal arrhythmia, heart failure, and ischemic mitral regurgitation can occur. Sudden death sometimes occurs in infancy. On the contrary, patients whose collateral pathways develop well can evade ischemic disorders and remain asymptomatic throughout childhood. However, as the shunt volume to the PA increases, myocardial perfusion decreases by the steal phenomenon and causes complications as similar to those during infancy. The latter form is classified into adult BWG syndrome and is a very rare disease [7]. Our patient had no noted physical abnormalities after birth and remained asymptomatic throughout her childhood. She had a history of three vaginal births without complications. However, she was aware of chest discomfort on exerting effort for the prior 3 years. When she rode a bicycle, she always felt chest discomfort. She previously changed her means of transportation from bicycle to motorcycle. However, she changed her means of transportation to bicycle for drinking alcohol during a party that day. The alcohol consumption and bicycle riding increased her heart load and exacerbated the ischemic disorder. Finally, lethal arrhythmia had been induced.

The risk of sudden death in patients with adult BWG syndrome is high, although it is commonly asymptomatic. In the report with 151 cases of adult BWG syndrome, 17% presented ventricular arrhythmia, syncope, or sudden cardiac death [8]. Surgical treatment to rebuild a dual coronary system is recommended regardless of symptom status at the time of diagnosis [9, 10]. In cases of lethal arrhythmia with organic origins such as old myocardial infarction, it is necessary to consider treatment adaptations such as an implanted cardioverter defibrillator (ICD) or catheter ablation [11, 12]. We recommended surgical treatment for our patient because we

thought that the threshold of the ischemic disorder had decreased, and the risk of complications and recurrence had increased. We performed a cardiac electrophysiology study to evaluate the reproducibility of lethal arrhythmia and found none. The risk of lethal arrhythmia recurrence was decreased due to improvement of the coronary blood flow abnormality and ischemia by surgical treatment. Finally, we judged that she was not a candidate for ICD or catheter ablation.

The earlier EMS arrives, the better the survival rate after OHCA [13]. The earlier defibrillation with AED is achieved, the more a patient's prognosis improves [14]. In addition, prognosis improves much more with than without BCPR [15]. In our case, EMS arrival time was 9 min, almost equal to the national average (8 min in 2015) [4], but 14 min passed prior to defibrillation and the return of spontaneous circulation. However, she was resuscitated without neurological impediments due to high-quality BCPR initiated immediately and continued by her two coworkers. This occurred for two reasons. First, her coworkers were nurses. Second, they had participated in a resuscitation training hosted by the city fire station and mastered the BCPR procedure several months prior. If there had been an AED nearby, they could have performed defibrillation sooner. In Japan and worldwide, the increase in educational interventions about resuscitation training significantly increases favorable neurological survival in OHCA cases treated with BCPR each year [16–18]. This case demonstrates the importance of education about BCPR.

Our patient returned to work as a nurse 6 months after surgery and continues to use her bicycle as a means of transportation today.

Conclusion

Here, we described a case of a woman with BWG syndrome who was resuscitated by BCPR without any subsequent neurological impediments. The complication of lethal arrhythmia by the steal phenomenon resulted in the CPA. This case demonstrates the importance and usefulness of BCPR training programs.

Authorship

HF: drafted the article. TW: substantially contributed to the conception and design. YS: substantially contributed to the conception and design. KI: revised critically for important intellectual content. TM: revised critically for important intellectual content. HI: revised critically for important intellectual content. KU: involved in acquisition of data, or analysis and interpretation of data. SI: involved in acquisition of data, or analysis and interpretation of data. KM: involved in acquisition of data, or analysis and interpretation of data. KT: revised critically for important intellectual content. YS: revised critically for important intellectual content. SH: made final approval of the version to be published.

Conflict of Interest

The authors have no conflicts of interest to disclose.

References

1. Hauser, M. 2005. Congenital anomalies of the coronary arteries. Heart 91:1240–1245.
2. Mazurak, M., and J. Kusa. 2014. The radiologist's tragedy, or Bland-White-Garland syndrome(BWGS). On the 80th anniversary of the first clinical description of ALCAPA (anomalous left coronary artery from the pulmonary artery). Kardiochirgia i Torakochirurgia Polska. 11:225–229.
3. Wesselhoeft, H., J. S. Fawcett, and A. L. Johnson. 1968. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. Circulation 38:403–425.
4. Fire and Disaster Management Agency. The present conditions of emergency and rescue. Fire and Disaster Management Agency. Available at http://www.fdma.go.jp/neuter/topics/kyukyukyujo_genkyo/h28/01_kyukyu.pdf (accessed 12 January 2017).
5. Blake, H. A., W. C. Manion, T. W. Mattingly, and G. Baroldi. 1964. Coronary artery anomalies. Circulation 30:927–940.
6. Keith, J. D. 1959. The anomalous origin of the left coronary artery from the pulmonary artery. Br. Heart. J. 21:149–161.
7. Edwards, J. E. 1964. The direction of blood flow in coronary arteries arising from the pulmonary trunk. Circulation 29:163–166.
8. Yau, J. M., R. Singh, E. J. Halpern, and D. Fischman. 2011. Anomalous origin of the left coronary artery from the pulmonary artery in adults: a comprehensive review of 151 adults cases and a new diagnosis in a 53-year-old woman. Clin. Cardiol. 34:204–210.
9. Manoly, I., D. Karangelis, N. Viola, and M. Haw. 2014. Repair of Bland-White-Garland syndrome via a modified technique. Tex. Heart Inst. J. 41:48–50.
10. Schwartz, M. L., R. A. Jonas, and S. D. Colan. 1997. Anomalous origin of left coronary artery from pulmonary artery: recovery of left ventricular function after dual coronary repair. J. Am. Coll. Cardiol. 30:547–553.
11. Parizek, P., L. Haman, J. Harrer, M. Tauchman, V. Rozsival, I. Varvarovsky, et al. 2010. Bland-White-Garland

syndrome in adults: sudden cardiac death as a first symptom and long-term follow-up after successful resuscitation and surgery. Europace 12:1338–1340.
12. Kristensen, T., K. F. Kofoed, S. Helqvist, M. Helvind, and L. Sondergaard. 2008. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) presenting with ventricular fibrillation in an adult: a case report. J. Cardiothoracic Surg. 3:33.
13. Rajan, S., M. Wissenberg, F. Folke, S. M. Hansen, T. A. Gerds, K. Kragholm, et al. 2016. Association of bystander cardiopulmonary resuscitation and survival according to ambulance response times after out-of-hospital cardiac arrest. Circulation 134:2095–2104.
14. Valenzuela, T. D., D. J. Roe, G. Nichol, L. L. Clark, D. W. Spaite, and R. G. Hardman. 2000. Outcomes of rapid defibrillation by security officers after cardiac arrest in casinos. N. Engl. J. Med. 343:1206–1209.
15. Hasselqvist-Ax, I., G. Riva, J. Herlitz, M. Rosenqvist, J. Hollenberg, P. Nordberg, et al. 2015. Early cardiopulmonary resuscitation in out-of-hospital cardiac arrest. N. Engl. J. Med. 372:2307–2315.
16. Kitamura, T., K. Kiyohara, T. Sakai, T. Matsuyama, T. Hatakeyama, T. Shimamoto, et al. 2016. Public-access defibrillation and out-of-hospital cardiac arrest in Japan. N. Engl. J. Med. 375:1649–1659.
17. Malta, H. C., K. Kragholm, D. A. Pearson, C. Tyson, L. Monk, B. Myers, et al. 2015. Association of bystander and first-responder intervention with survival after out-of-hospital cardiac arrest in North Carolina, 2010-2013. JAMA 314:255–264.
18. Kitamura, T., T. Iwami, T. Kawamura, M. Nitta, K. Nagao, H. Nonogi, et al. 2012. Nationwide improvements in survival from out-of-hospital cardiac arrest in Japan. Circulation 126:2834–2843.

