Anorexia Nervosa in a Postoperative Patient With Ebstein Anomaly

Sato Kengo (tgv_thalys@hotmail.com)
Jichi Medical University
Watanabe Ryosuke
Jichi Medical University
Okada Tsuyoshi
Jichi Medical University
Nishiyori Yasushi
Jichi Medical University
Kobayashi Toshiyuki
Jichi Medical University
Suda Shiro
Jichi Medical University

Research Article

Keywords: Adult congenital heart disease, Anorexia nervosa, Ebstein's anomaly, Fontan procedure

Posted Date: June 29th, 2021

DOI: https://doi.org/10.21203/rs.3.rs-616147/v1

Abstract

Background: Along with the improved life prognosis of patients with congenital heart disease, their diverse complications have come under scrutiny. Due to the various medical restrictions attendant on their upbringing, patients with congenital heart disease often have coexisting mental disorders. However, reports of patients with congenital heart disease and coexisting eating disorders are very rare. In this paper the authors report the case of a patient who developed anorexia nervosa following surgery for Ebstein's anomaly.

Case Presentation: A 21-year-old female with Ebstein's anomaly who underwent a Fontan surgery was transferred to our institution with suspected anorexia nervosa after a more than 2-year intermittent stay in a medical hospital for decreased appetite. Initially, she did not express a desire to lose weight or a fear of obesity, and we suspected that she was suffering from appetite loss due to a physical condition associated with the Fontan circulation. But gradually the pathology of the eating disorder became apparent.

Conclusions: Patients with congenital heart disease are more likely to have a psychological background as well as physical problems that might contribute to eating disorders. Indeed, it is unclear why there are not more cases of eating disorders in association with congenital heart diseases.

Background

Due to the development of new medical technologies, approximately 90% of children with congenital heart disease can survive into adolescence and adulthood [1]. In 2007, there were approximately 410,000 such patients in Japan and the number has been increasing at a rate of 10,000 per year in that country [2]. However, in adulthood, these patients have to face not only problems directly related to the functional status of the heart, such as limited life expectancy, restricted degree of cardiac function, rehospitalization, reoperation, postoperative residuals, and psychological burden, but also problems related to education, employment, marriage, childbirth, and social security (insurance, pensions, certification of disability, medical benefits, rehabilitation medicine, etc.) [3, 4]. Their medical and social difficulties are likely to be a precipitating factor for mental disorders.

In this paper, the authors report a case of anorexia nervosa in a patient with Ebstein's anomaly, a severe congenital heart disease, who underwent Fontan surgery. Perhaps because there are few reports on the combination of congenital heart disease and anorexia nervosa, the question of whether the loss of appetite originates from a physical condition or an eating disorder can confuse clinicians.

In order to avoid identifying our patient, some details have been slightly altered without impacting the argument. The patient gave verbal consent for this report.

Case Presentation

The patient was a 21-year-old female at the time of her first visit to our department. Her father has major depressive disorder. She was born as the first child of two siblings. She was diagnosed with Ebstein's anomaly and underwent a Blalock-Taussig shunting at 1 month of age. At the age of 8 years, she developed a fear of taking food and was temporarily unable to eat. After undergoing counseling sessions she recovered from the condition. At the age of 9, she underwent a Fontan surgery, and her physical condition improved considerably. Although she had to move around the country for hospital visits, she was able to go to school with the support of her friends.

In April of her 18th year, after graduating from high school, she got a job as an office worker at an industrial company. Her weight at this point was 45 kg, with a body mass index (BMI) of 18.5. At the start of her employment, she engaged in her work without difficulty, but 6 months later, she was transferred to a new position against her will. The new position required her to engage in strenuous work, and she became fatigued, complained of frequent stomachaches, and lost her appetite. In February at the age of 19, during a regular visit to the cardiovascular surgery clinic, hypotension and hypokalemia were noted, and she was referred to the internal medicine ward of a general hospital for admission. Her hypotension and hypokalemia were ameliorated with treatment focusing on fluid supplementation, but her weight loss was not fully recovered. She was discharged from the hospital at a weight of 40 kg and returned to work, but her physical condition again worsened due to decreased food intake, and she was readmitted to the hospital. Over the subsequent 2 years, she was repeatedly hospitalized a total of 13 times in the same way.

Since the development of an eating disorder was suspected, she was referred to the psychiatric clinic of the hospital and olanzapine 2.5 mg was started, but it was soon discontinued due to water intoxication. She was also referred to the psychiatric department of another hospital and started diazepam 10 mg, but there was no effect. In March of her 20th year, she was admitted to the internal medicine ward of the hospital for the 14^{th} time due to a deteriorating physical condition. She was unable to discuss her concerns about food intake with the staff due to denial and loss of insight, and she exhibited food refusal behaviors, such as spitting her food out..

In June, she was referred and transferred to our department for treatment of anorexia nervosa. She asserted that she did not want to lose weight and that the thought of losing weight depressed her. She weighed 31.9 kg, with a BMI of 13.3 kg/m^2. There was no limb edema. A head CT scan showed no abnormal findings. The WAIS-III performed in the third month of hospitalization showed a full scale IQ of 74, with a verbal IQ of 82 and motor IQ of 71, which was a borderline intelligence level. We started her on a diet of 1150 kcal plus 400 kcal of nutritional supplements. After admission, she took all meals, but on the 5^{th} day, she was still 32 kg, and we considered the possibility that she was vomiting. She said "I never feel sick to my stomach. I don't know why my weight isn't increasing." She began complaining of nausea regularly, but only rarely vomited. Since her weight did not increase commensurate with the amount of food intake during the first month of admission, we suspected that physical factors related to her heart disease were suppressing her weight gain. In fact, an abdominal CT scan revealed a large amount of ascites and intestinal edema (Figures 1 and 2). There were neither significant changes in her blood tests

nor decrease in protein or albumin. In the opinion of the pediatric cardiologist, her massive ascites and intestinal edema might have come from a special form of circulation called Fontan circulation, which is characterized by elevated venous pressure. Thus, we considered the possibility that she was unable to consume food because of a fear that eating would aggravate her gastrointestinal symptoms. In addition, because there was a possibility of protein leak gastroenteropathy as a complication of the Fontan circulation, 99mTc-HSA-D gastrointestinal scintigraphy was performed on the 42nd day, which revealed that there were no findings suggestive of this disease. Consequently, the ascites was considered to be due to hypoproteinemia associated with inadequate dietary intake. On the 49th day, her weight dropped to the 30 kg range, and nasogastric tube feeding was required starting at 1600 kcal. Her weight gradually increased but stagnated on the 71st day, and her total nutrition was increased to 2,000 kcal. On the 89th day her weight reached 38.3 kg. A CT scan performed on the 91st day showed that the ascites had almost disappeared. She was able to take almost 100% of her food orally, without vomiting, so she was discharged on the 150th day.

After one month of stable food intake and weight maintenance, at the end of November of the same year, she started vomiting again, and the vomiting became more frequent. Her weight drastically decreased, and by December, dropped to 31 kg. Her appetite was markedly decreased and her nausea worsened when she ate, so she had to be admitted to our hospital for the second time. Her weight at admission was 29.4 kg, with a BMI of 12.2 kg/m^2. Given that nausea caused by intestinal edema was a factor in the poor feeding, early improvement of nutritional status was considered to be the key to her treatment. Although she accepted the nasogastric tube feeding well at first, when we proposed a specific target weight of 40 kg, she expressed her resistance to weight gain, saying, "I think that is a bit much." Nevertheless, on the 24th day, her weight had recovered to 33 kg, and she expressed a desire to eat by mouth. So, the nasogastric tube was removed. As her weight reached 35 kg, she began to eat less. She also started to show overactivity, moving around the ward frequently. At this point, we concluded that her weight gain stagnation was probably due to vomiting in secret. She also told other patients that she had thrown up so much that she had lost too much weight, indicating that she was deliberately trying to control her weight. However, she did not refuse to gain weight and her acceptance of medical treatment remained good.

On the 101st day, she exceeded 39 kg and began to express her fear of weight gain, and demanded to be discharged from the hospital, stating "I am afraid that if I exceed 40 kg, that will be too heavy for me, and my weight gain might spiral out of control." As her weight gain was sluggish at around 40 kg, we respected her wishes and discharged her on the 191st day. However, soon after that she lost weight again and was readmitted to the hospital, and now, about 4 years after her first hospitalization, she is under her fifth admission at our hospital.

Over the course of 4 years, she was out of the hospital for only 99 days in total. Including the time spent at the previous hospital, she has been hospitalized for over 6 years. Her attempt to control her weight remain serious enough that she cannot be allowed to be discharged from the hospital, although the restrictions on her activities in the hospital environment are eased when she stops vomiting. The family

cannot hide their anxiety about caring for her at home with her existing medical and psychiatric disabilities, and they are searching for a next step, such as transferring her to a psychiatric hospital in anticipation of long-term inpatient care.

Discussion

Ebstein's anomaly is a congenital heart disease in which one or two valve leaflets of the tricuspid valve are displaced into the right ventricle, resulting in obstruction of tricuspid valve closure and severe regurgitation. The final functional repair procedure is the Fontan surgery, in which venous blood from the body circulation flows directly to the pulmonary artery, i.e., the superior and inferior vena cava are anastomosed directly to the pulmonary artery. With advances in the techniques of Fontan surgery, the quality of life and life prognosis of patients with single ventricle circulation have been remarkably improved, with a reported survival rate of 94% at 10 years after surgery [5]. However, Goldberg cautions that the Fontan circulation is an "artificially created chronic heart failure circulation" with high central venous pressure and low cardiac output, which is different from normal circulatory physiology [6]. Furthermore, although the precise mechanism is unknown, high venous pressure might cause complications closely related to dietary intake, such as protein leak gastroenteropathy and liver fibrosis [7,8].

In the present case, there were no typical symptoms of anorexia nervosa upon admission to our hospital. It seemed that if the patient had an eating disorder, it would fall under the category of avoidance/restrictive food intake disorder. Indeed, the fact that she did not resist the tube and intravenous feedings at her previous hospital was not typical of anorexia nervosa. It was only gradually after the second hospitalization that the pathology of anorexia nervosa became apparent. As Nakai et al. [9] pointed out, there are cases in which a diagnosis of avoidance/restrictive food intake disorder is initially made, but the pathology of anorexia nervosa becomes apparent later, in parallel with this case.

As far as we know, there are no published reports of cases of congenital heart disease coexisting with eating disorders. On the other hand, there have been many reports of Turner's syndrome, which is associated with a high rate of aortic stenosis, concomitant with anorexia nervosa. It is thought that hormone therapy and the psychosocial positioning of the patient as having Turner's syndrome are related to the development of anorexia nervosa [10]. Similarly, in the present case, we suspect that the combination of congenital heart disease and an eating disorder was not a simple coincidence. Wray et al. [11] stated that patients with congenital heart disease are significantly delayed in social maturation, and that one of the triggers of eating disorders is a lack of social contact due to a dependent lifestyle caused by parental overprotection. In this case, the patient spent more time in the hospital than at home due to frequent hospitalizations and multistep surgeries since infancy, which resulted in social isolation and insufficient contact with others. It can be said that the parents devoted their energies to her treatment, diligently visiting hospitals all over the country, but as she had a disability, the parents had no choice but to be protective, which may have led to overprotection and its associated affects on mental development.

In addition, the protective environment of the hospital might have increased her dependency. It is likely that this patient, who was late in maturing socially, was frustrated by the ordeal of finding a job, and when she retreated to the hospital, she re-experienced a familiar and protective environment she had known since childhood. It has been pointed out that adult patients with congenital heart disease have a high incidence of neurocognitive disorders, which can lead to poor social adjustment [12]. The intellectual level of the patient in this case was borderline, which may also have played a role in her deficiency of social adjustment. The background to the development of eating disorders is diverse and cannot be summarized in a single sentence, but it is often pointed out that the relationship with parents and self-esteem play a role [13,14]. The presented case suggests that difficulties in social maturation in children with congenital heart disease may lead to the pathology of eating disorders. Indeed, it is unclear why there are not more cases of eating disorders in patients with congenital heart diseases. Nevertheless, when this complication occurs, it becomes difficult to diagnose whether the loss of appetite is due to a physical condition or an eating disorder.

Our patient's ability to express emotions was poor, and although she often paints pictures as a recreational activity on the ward, her paintings are not highly expressive and do not provide clues to her feelings. Therefore, it is not easy to know her inner world, but it seems that many of her comments are related to immediate situations or brief emotions and lack a long-term perspective. She is unable to find a way to mediate between her feelings of not wanting to be restricted in her activities, not wanting to gain weight, and not wanting to go to work. For the time being, she is unable to maintain her weight without physical danger unless she is in an inpatient environment. She requires long-term inpatient treatment which will allow her to explore her own path to self-realization.

Conclusion

We have reported a case of anorexia nervosa in a postoperative patient with Ebstein anomaly. A combination of parental protective measures due to her congenital heart disease, increased dependence due to prolonged hospitalization, neurocognitive dysfunction due to the primary disease, and gastrointestinal symptoms due to Fontan circulation were considered to have resulted in anorexia nervosa as a means of avoiding social participation. In other words, the social difficulties of adult patients with congenital heart disease may have taken the form of an eating disorder.

Abbreviations

Not applicable.

Declarations

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Authors' contributions

Kengo Sato conceived of the case report and wrote the manuscript. All authors read, edited, reviewed, and approved the final manuscript.

Ethics approval and consent to participate

The patient consented to the use of anonimized medical data, including diagnostic images, in our paper.

Consent for publication

We give consent for the publication of our paper.

Availability of data and materials

Not applicable.

Competing interests

We declare no competing interests.

Funding

This research received no grant or funding from any source.

Acknowledgements

Not applicable.

Author detail

Department of Psychiatry, Jichi Medical University, Tochigi, Japan

References

1. Moons P, Bovijn L, Budts W, et al. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. Circulation. 2010;122:2264–72.

2. Motoki H. Management of congenital heart disease in adult. Shinshu Igaku Zasshi (Shinshu Medical Journal). 2014;62:285–94. (in Japanese)

3. Moller JH, Taubert KA, Allen HD, et al. Cardiovascular health and disease in children: Current status. Circulation. 1994;89:923–30.

4. Perloff JK, Warnes CA. Challenges posed by adults with repaired congenital heart disease. Circulation. 2001;103:2637–43.

5. Nakano T, Kado H, Tachibana T, et al. Excellent midterm outcome of extracardiac conduit total cavopulmonary connection: Results of 126 cases. Ann Thorac Surg 2007;84:1619–25.

6. Goldberg DJ. The Fontan operation. Improved outcomes, uncertain future. J Am Coll Cardiol. 2015;66(15):1711–3.

7. Mertens L, Hagler DJ, Sauer U, et al. Protein-losing enteropathy after the Fontan operation: An international multicenter study. J Thorac Cardiovasc Surg 1998;115:1063–73.

8. Feldt R, Driscoll DJ, Offord KP, et al. Protein-losing enteropathy after the Fontan operation. J Thorac Cardiovasc Surg 1996;112:672–80.

9. Nakai Y, Nin K, Teramukai S, Taniguchi A, Fukushima M, Wonderlich SA. Typical and atypical anorexia nervosa in a Japanese sample. International Journal of Eating Disorders,2014;47:130–7.

10. Darby PL, Garfinkel PE, Vale JM, et al. Anorexia nervosa and 'Turner syndrome': cause or coincidence? Psychol Med. 1988;11:141–145.

11. Wray J, Sensky T. How does the intervention of cardiac surgery affect the self-perception of children with congenital heart disease? Child: Care, Health and Development. 1998;24:57–72.

12. Cohen S, Earing M. Neurocognitive impairment and its long-term impact on adults with congenital heart disease. Prog. Cardiovasc. Dis. 2018;61:287–93.

13. Bruch H. The Golden Cage: The Enigma of Anorexia Nervosa. Cambridge, MA: Harvard University Press; 1978.

14. Minuchin S. Psychosomatic Families: Anorexia Nervosa in Context. Cambridge, MA: Harvard University Press; 1978.

Figures

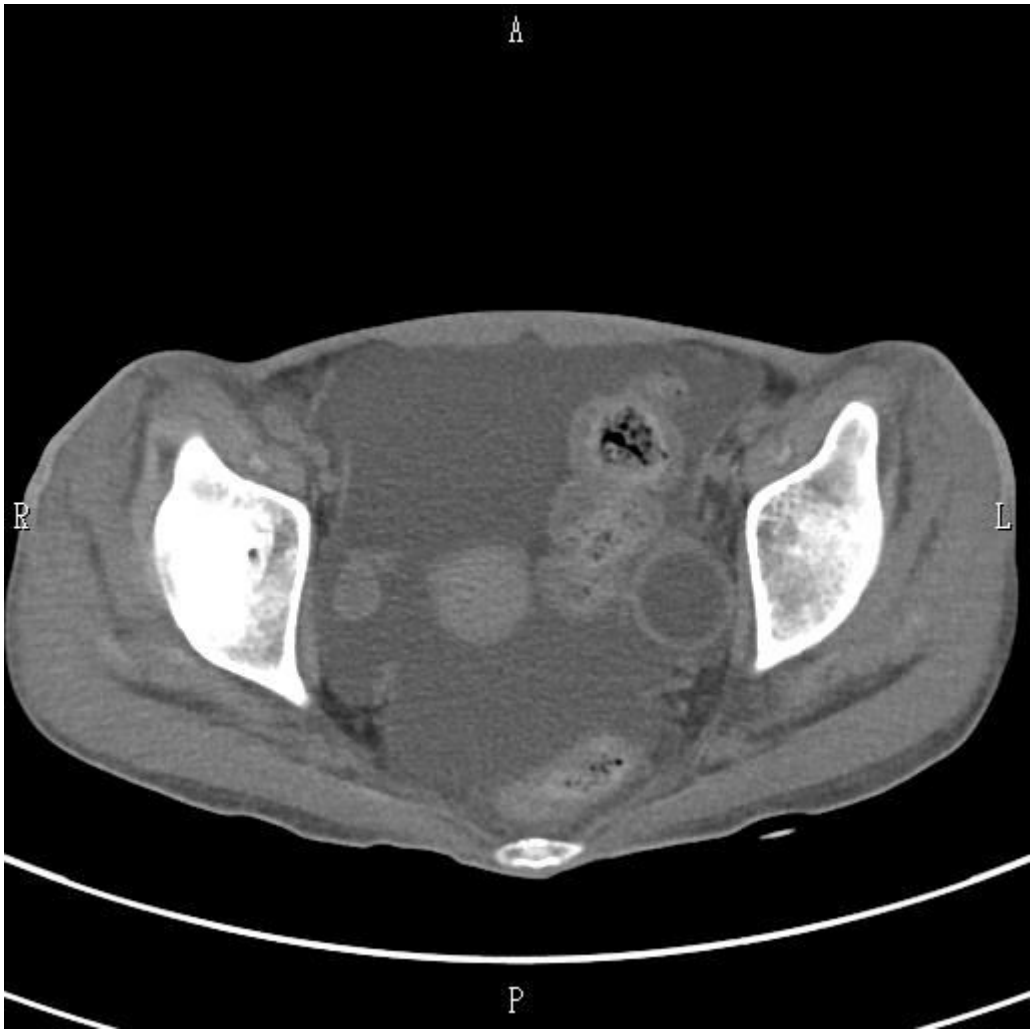

Figure 1

CT abdomen (transverse view). A large amount of ascites was apparent around the intestine.

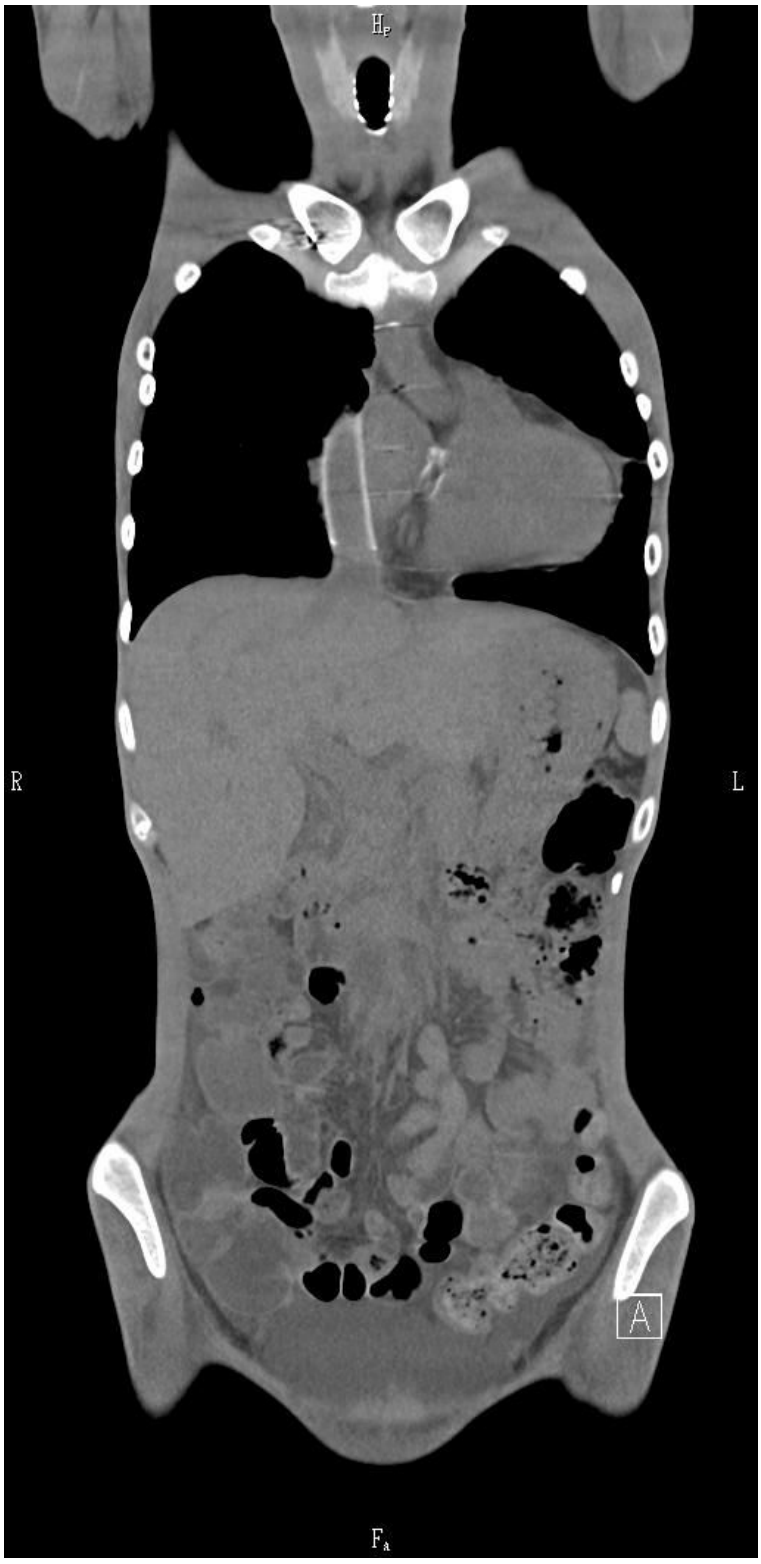

Figure 2

CT abdomen (longitudinal view). There is thickening of the intestinal wall, suggesting intestinal edema.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- care.pdf