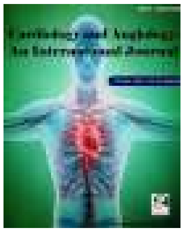

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# Athlete's Heart with a Systemic Right Ventricle?

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***Authors' contributions***

*This work was carried out in collaboration between all authors. Authors JM and AH were responsible for conception and design of the case study. Author JM conducted the cardiopulmonary exercise test, sampled and analysis the data and drafted the manuscript. Author SF conducted the magnet resonance imaging. All authors gave important input for revising the manuscript and read and approved the final version of the manuscript.*

***Case Study***

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## ABSTRACT

**Objective:** High load of regularly vigorous exercise leads to multiple physiological adaptations. The major cardiovascular effects are hypertrophy and dilation, predominantly of the left ventricle, and bradycardia. However, there are no reports on an athlete's heart in a systemic right ventricle.

**Subject:** We report on a 23 year old male endurance athlete (177cm, 69kg) with a systemic subaortic right ventricle after atrial redirection (Senning procedure) for simple transposition of the great arteries in infancy. Albeit medical doctors had imposed activity restriction to him, he has lead an active lifestyle from early childhood on, intensifying his sport activities over the years especially in cycling and running to a training volume of about 10 hours per week in winter and about 15 hours per week in summer. In 2009 he performed 1:50h on the half marathon distance. In 2013 he finished his first Marathon in 4:34h.

**Results:** Cardiopulmonary exercise testing revealed a maximum oxygen uptake of 52.3ml/min/kg and a peak work load of 353 Watt, corresponding to 5.1Watt per kilogram body mass. Cardiovascular Magnetic Resonance showed a cardiac index of 2.9ml/min/m², a tricuspid regurgitation fraction of 4%, and a systemic right ventricle end-diastolic volume of 109ml/m² with an ejection fraction of 53%.

**Conclusions:** With regular exercise training a systemic right ventricle can become very efficient comparable to healthy amateur athletes.

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## 1. INTRODUCTION

High load of regular vigorous exercise leads to multiple physiological adaptations. The major cardiovascular effects are hypertrophy and dilation of the left ventricle, as well as bradycardia. This is known as the characteristical clinical picture of “athlete’s heart” [1,2]. Recent studies suggest that there is also an increase in right ventricular diameters in endurance athletes [3].

In patients after atrial redirection for transposition of the great arteries there is a systemic right ventricle with an outflow through the aortic valve into the systemic circulation. Since this ventricle has to deal with the systemic blood pressure and systemic resistance, there is also a morphologic adaption of the right ventricle, maybe comparable to athlete’s heart.

However, those patients often suffer from exercise intolerance [4,5] and have a reduced ability to increase stroke volume under exercise [6]. Reasons for this are complex but include concerns like functional obstruction by the incompliant atrial baffles, dysfunction of the right ventricle that has to act like a systemic left ventricle, or tricuspid regurgitation. The low heart rate response to exercise might also contribute [4].

However, it should be considered that exercise intolerance could also be the result of exercise restrictions given to those patients and their parents from medical staff from early childhood on.

## 2. CASE REPORT

We report on a 23 year old male athlete (177cm, 69kg) with simple transposition of the great arteries. This is a congenital heart defect with an aorta arising anteriorly from the right ventricle and a pulmonary artery with an origin posteriorly from the left ventricle. Initial atrial switch operation (Senning procedure) was performed at the 23rd day of life, redirecting the systemic venous blood return from the caval veins to the mitral valve and redirecting the pulmonary venous return to the tricuspid valve. This procedure results in a normal sequential blood flow, but the left ventricle is serving the pulmonary vascular bed and the right ventricle is serving the systemic circulation. That leads to right ventricular hypertrophy since the right ventricle has to cope with the systemic resistance. Due to the changing geometry, tricuspid regurgitation is a common sequel in the long-term and right ventricular failure also becomes present very often beyond the age of 30.

Since then no further surgical intervention was necessary. No long-term medication was reported. He was employed full-time in an office. Echocardiography revealed a good function of both ventricles, only trivial tricuspid valve regurgitation, and no signs of any valve stenosis.

Albeit medical doctors impose activity restriction to him, he had lead an active lifestyle from early childhood on and participated in school sport and leisure sport activities with his healthy peers. During the last couple of years he intensified his sport activities especially in cycling and running to a training volume of about 10 hours per week in winter and about 15 hours per week in summer. During spring and summer his mileage was about 5000 cycling kilometres, whereas in fall/winter his training focused more on running performance. He had participated in multiple competitive sport events like bicycle marathons and running competitions. In 2009 he performed 1:50h on the half Marathon distance, in 2013 he finished his first Marathon in 4:34h.

During an annual cardiologic examination he performed a cardiopulmonary exercise test (CPET) on a bicycle ergometer in upright position as previously described [7] with a ramp wise increase of the load with 40Watt per Minute. Afterwards he underwent cardiovascular magnetic resonance (CMR) as previously described [6].

During exercise, heart rate increases from 70 to 172 beats per minute as well as blood pressure from 128/85mmHg to 180/87mmHg at peak exercise. There was no drop in oxygen saturation throughout the test. Achieved maximum work load was 353 Watt according to 5.1 Watt per kilogram body mass. Maximal oxygen uptake ( $\dot{V}O_2$ max) was 3.66 litres per minute corresponding to 52.3ml/min/kg, respectively. CPET was performed till muscular exhaustion showing a leveling off in oxygen uptake and a respiratory exchange ratio of 1.15.

CMR showed the typical right heart dilation and hypertrophy as it is seen in patients with a systemic right ventricle (Table 1, online Supplement). Cardiac index was 2.9ml/min/m² and the Tricuspid valve regurgitation fraction was 4%.

**Table 1. Right ventricular parameters derived from magnet resonance imaging parameters in comparison to a TGA cohort from Fratz et al. [6]**

| | Study subject | Reference values |
|---|---|---|
| End-diastolic volume index (ml/m$^2$) | 109 | 98 (59-198) |
| End-systolic volume index (ml/m$^2$) | 101 | 54 (21-159) |
| Stroke volume index (ml/m$^2$) | 57 | 45 (34-58) |
| Ejection fraction (%) | 53 | 44 (20-75) |

## 3. DISCUSSION

This case report demonstrates that also a systemic right ventricle can be very efficient. A systemic right ventricle is able to provide a good adaption to a long-term endurance exercise training resulting in a performance comparable to healthy amateur athletes. Moreover, it outlines the beneficial effect of regular endurance exercise on maximal oxygen uptake and on the hemodynamic situation even in this patient with complex CHD and a systemic right ventricle.

Even if assuming an individual with a perfect surgical outcome and no present residuals, the $\dot{V}O_2$ max of 52.3ml/min/kg exceeded comparative values from other TGA cohort studies [4,5] by more than the double. In addition, also the work load of 353Watt (5.1Watt/kg body mass), respectively, is comparable to healthy amateur athletes of similar age.

As expected, CMR revealed a good function of both ventricles. However, we only found the same morphological adaption and volumes as previously reported in a cohort of untrained individuals with TGA after the Senning procedure [6]. Right heart hypertrophy is the leading morphological adaption in patients with TGA as the right ventricle had to act as the systemic ventricle with the whole systemic afterload after the atrial redirection in infancy. We speculate that this early adaption might overlay the physical adaption from regular endurance sport in our highly trained subject.

Thus, the high performance of our individual must be referred to other physiological modulations of continuous endurance training. Increase in the number and size of mitochondria, a slower utilization of muscle glycogen and blood glucose, a greater reliance on fat oxidation, and less lactate production during exercise also play an important role in the ability to perform prolonged strenuous exercise apart from the increased cardiac output of an "athlete heart" [8]. All those single mechanisms lead to improved global exercise performance also in patients with CHD since contemporary studies [9-11] have reported on the association of exercise performance and regular physical activity.

The pioneering training study from Fredriksen et al. [10] initially outlined the benefit of a heart rate based training program and also Dua et al. [9] showed that already regular walking exercise had measurable effects on oxygen uptake. In a more recent study we have shown that there is a positive relation between the amount of at least moderate activity per day and peak oxygen uptake [11]. Thus, in this special case an excellent surgical result combined with a high amount of exercise, structured in a seasonal training schedule, might be the key for the outstanding exercise performance in a patient with a systemic right ventricle.

However, our initial question whether subjects with a systemic right ventricle can develop an athlete's heart could not be clarified. Morphologic adaptations already begin after the surgical repair. The right ventricle adapts to its new function and this might overlay the latter adaption from regular endurance training. Nevertheless this individual depicts that patients with TGA after atrial redirection can merit tremendously from endurance exercise training.

## 4. CONCLUSION

With regular exercise training also a systemic right ventricle can become very efficient comparable to healthy amateur athletes.

## CONSENT

All authors declare that written informed consent was obtained from the patient for publication.

## ETHICAL APPROVAL

The study was prospectively designed and in accordance with the declaration of Helsinki (revision 2008).

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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