

A cardiac surgeon, his way to holiness, and his heritage – the 80th anniversary of Giancarlo Rastelli's birthday and the 45th anniversary of the first Rastelli procedure

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Abstract

Cardiac surgeon Giancarlo Rastelli was born 80 years ago in Pescara, Italy. His name was immortalized in such eponyms as the Rastelli classification (classification of atrioventricular septal defects) and the Rastelli procedure (among other applications, used for the treatment of transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction). Giancarlo Rastelli died in 1970 at the age of 36. His beatification has been in progress at the Vatican since 2005. This paper presents the current view on the Rastelli classification, the problems related to the Rastelli procedure, as well as alternative procedures applied in specific cases, i.e. the Nikaidoh, REV, and Metras procedures.

Key words: Rastelli procedure, Rastelli classification, history of medicine.

Streszczenie

Giancarlo Rastelli urodził się 80 lat temu w miejscowości Pescara we Włoszech. Jego nazwisko zostało nieśmiertelnie w postaci eponimów określających klasyfikację ubytku przegrody przedsionkowo-komorowej (klasyfikacja Rastelliego) oraz technikę zabiegu operacyjnego stosowanego m.in. w przełożeniu wielkich pni tętniczych z ubytkiem przegrody międzykomorowej i zwężeniem drogi odpływu lewej komory (operacja metodą Rastelliego). Giancarlo Rastelli zmarł w 1970 r. w wieku 36 lat. Od 2005 r. w Watykanie trwa jego proces beatyfikacyjny. W opracowaniu przedstawiono aktualne spojrzenie na klasyfikację Rastelliego, jak również problemy związane z operacją metodą Rastelliego oraz alternatywne metody leczenia stosowane w poszczególnych przypadkach, tj. metodą Nikaidoh, REV i Metras.

Słowa kluczowe: operacja Rastelliego, klasyfikacja Rastelliego, historia medycyny.

Introduction

In 1969, a 12-year old boy, Vincenzo Ferrante, arrived in the USA (Houston, Texas) with his parents. He was supposed to be operated on by surgeons from Texas, since the previous surgical procedure that he had undergone (atrioseptectomy, i.e. the Blalock-Hanlon procedure) 3 years before was performed there as well. Unfortunately, it turned out that the level of the defect's complexity (transposition of the great arteries with ventricular septal defect and pulmonary valve stenosis) disqualified the patient from further treatment. The desperate parents turned for help to Doctor Giancarlo Rastelli. The cardiac surgeon, himself

struggling with Hodgkin's lymphoma for almost a year, decided to diligently treat his compatriot. The boy underwent two operations at the Mayo Clinic in Rochester in November 1969. The first was the Rastelli procedure, but, unfortunately, reoperation was necessary due to the recanalization of ventricular septal defect. The condition of the young Italian was improving each day, while the health of Doctor Giancarlo Rastelli was gradually deteriorating. In spite of this, the physician visited his patient every day. He even spent Christmas Eve next to his bed, because the 12-year-old had to stay at the clinic for the holidays. Vincenzo Ferrante left the hospital after several weeks and returned to

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Italy with his parents. He went to university, became an engineer, working in Naples. Giancarlo Rastelli died less than a month after the boy was discharged from the hospital on February 2, 1970. He was 36 years old. He died at the peak of his professional career. The process of his beatification in the Vatican has lasted since 2005... [1, 2].

Who was the man whom modern doctors working with congenital heart defects usually associate with such eponyms as the Rastelli classification and the Rastelli procedure? What were the extraordinary circumstances that may result in the unusual event that for the first time in history the words “blessed” and, later on, “saint” may appear before the name of a cardiac surgeon?

Giancarlo Rastelli was born 80 years ago on June 25, 1933 in the town of Pescara, Italy. His father, Vito Rastelli, was a journalist for a local newspaper, and his mother, Luisa Bianchi, worked as a primary school teacher. Giancarlo also had a younger sister, Rosangela. His uncle (Lino Rastelli) was a famous pianist. Having graduated from high school at the age of 17, Giancarlo was accepted into the medical faculty at the University of Parma. He developed an interest in surgery during the third year of his studies. Later on, he narrowed his focus to cardiovascular surgery. He graduated from the University of Parma with distinction in 1957, receiving a *cum laude* diploma for his work entitled: *Changes in the ATPase activity of the myocardium during deep hypothermia*. Three years later, Giancarlo Rastelli became a NATO scholarship holder, which enabled him to choose one of several centers in the USA. He chose the prestigious Mayo Clinic in Rochester, where one of the pioneers of cardiac surgery, John Kirklin (1917-2004), worked. Initially, Rastelli focused on invasive cardiac surgery, exploring the pathomorphology and pathophysiology of the circulatory system at the laboratory of cardiac catheterization and angiocardiology, supervised by Jeremy Swan. In this manner he established the basis for his experiments in cardiac surgery. In 1962, Rastelli started assisting at his first cardiac surgery procedures. John Kirklin – his mentor and teacher – wrote years later: *I personally have learned many things from Dr. Rastelli. In the numerous scientific projects that we worked on together, new knowledge was developed which has been of both practical and theoretical importance to me* [1]. Rastelli was especially interested in one congenital heart defect: atrioventricular septal defect (AVSD), the correction of which was associated with a very high mortality rate. As a result of his pathomorphological research conducted on a large number of dissected hearts, in 1967 he published a new classification of AVSDs [1-4].

The Rastelli classification

The Rastelli classification is based on the morphology of the superior bridging leaflet of the common atrioventricular canal and its relation to the interventricular septum and the papillary muscle of the right ventricle.

In type A (50-70% of all AVSD cases), the superior bridging leaflet is symmetrically medially divided along the line of the interventricular septum, with its two parts, left and

right, attached with multiple chordae tendineae to the interventricular septum in their respective ventricles. The left side of the common valve is displaced towards the apex and forms a common annulus with the right side. Type A often coexists with Down syndrome and is usually associated with pulmonary hypertension. In type B (3% – the rarest of the types), the superior leaflet is asymmetrically divided in such a way that its left component is partially displaced above the right ventricle, and the chordae tendineae cross the ventricular septal defect reaching the papillary muscle of the right ventricle. It is usually accompanied by the lack of one left ventricular papillary muscle, and the mitral part of the common valve may then form a parachute valve. Type B is often associated with unbalanced AVSD with right ventricular dominance. Type C of AVSD (30%) is characterized by an extreme displacement of the left superior leaflet above the right ventricle, so that it is freely located above the ventricular septal defect, and the chordae tendineae branching from it are attached to the papillary muscles of both ventricles. This form often coexists with heterotaxy syndrome and with conal defects [5-7].

In 1967, Rastelli *et al.* published a paper presenting the experience of the Mayo Clinic in operating on patients with complete AVSDs (38 patients, 1955-1967). Thanks to the better understanding of this defect and the adjustment of treatment to the new classification, the authors managed to reduce mortality from 60% (operations before 1964) to 20%. They also concluded that the division of AVSD into three anatomical subtypes enabled a better understanding of the nature of this defect and contributed to the improvement of surgical treatment outcomes [8]. It was very important for Rastelli to have his book-monograph devoted to AVSD published. He had to work against time because of the progression of his disease. Unfortunately, he did not manage to finish his work and left this task for others to complete. The monograph was finally published in 1976 [2].

Today, the Rastelli classification is no longer as useful as when it was first created. It did not take into account the numerous anatomical variations of AVSD, because of which some surgeons rejected it altogether. Some believe that the degree of leaflet restriction (tethering) is more relevant; therefore, in their opinion, the Rastelli classification today is only of historical importance [6]. Others claim, however, that despite the diversity of the anatomical variants of AVSD, the classic division of Rastelli still remains a very useful, clear, and effective tool in surgical practice [9].

There is also one more eponym related to AVSD containing the name of Giancarlo Rastelli, referring to a surgical technique used for repairing anomalies typical of this defect. This method, the Rastelli procedure, was implemented in 1967. It was based on the use of a single patch covering both the atrial and ventricular septal defects. The method was later modified: Dacron or Gore-Tex patches were used for closing ventricular septal defects, while pericardial patches were employed for repairing atrial septal defects. Currently, in selected cases, a modified version

of the single-patch technique is used, in which the valve components are attached to the apex of the interventricular septum. In 2011, the results of a study by French authors were published, which included the analysis of 107 infants below the age of 1 with complete AVSD, operated on between 1985 and 2006 in Marseille. The initial total operative mortality rate of 13% dropped to 4% within the last 10 years. The authors of the cited work stated that the Rastelli procedure for repairing AVSD is safe and repeatable, and it provides satisfactory results [9].

The Rastelli procedure

In the years 1962-1964, Giancarlo Rastelli conducted surgical experiments at the experimental laboratory of the Mayo Clinic. He was initially interested in persistent truncus arteriosus, a heart defect inoperable at that time. He attempted to close the ventricular septal defect and use a homograft to connect the right ventricle to the pulmonary trunk separated from the common trunk. His successful experiments encouraged Dwight McGoon to perform the first operation using this technique in 1967. The use of this innovative solution, and the application of a valve homograft connecting the right ventricle with the pulmonary trunk combined with the introduction of a patch directing blood flow from the left ventricle through the ventricular septal defect to the aorta, became the basis for the method currently known as the Rastelli procedure. Originally, this procedure was used in treating transposition of the great arteries (TGA) with ventricular septal defect (VSD) and left ventricular outflow tract obstruction (LVOTO). The first surgery using the Rastelli procedure to repair this complex defect was performed at the Mayo Clinic by Robert Wallace on July 26, 1968, precisely 45 years ago. The method was utilized a year before during the world's first successful repair of persistent truncus arteriosus (also at the Mayo Clinic, Dwight McGoon) [1, 2, 10, 11].

The Rastelli procedure is also currently used in other heart defects, including double outlet right ventricle with left ventricular outflow tract obstruction or atresia. Despite the fact that almost half a century has passed since the first Rastelli procedure was performed, the surgical treatment of patients with TGA, VSD, and LVOTO continues to pose a challenge. The long-term results of the Rastelli procedure remain a relevant object of study. The synthetic material used in LVOTO reconstruction does not have any growth potential. Moreover, the risk of LVOTO increases with time because of the natural tendency of VSD to gradually close, resulting in dysfunction of the left ventricle and arrhythmias. In order to avoid LVOTO, modifications were introduced consisting in the excision of the conal septum and widening of the VSD before establishing an outflow tract from the left ventricle to the aorta by means of a patch. This improved the early and mid-term survival after the Rastelli procedure. The late stenosis of the right ventricle to pulmonary artery (RV-PA) conduit requiring reintervention still remains a significant problem. Initially, the treatment results of patients after the Rastelli procedure were far from opti-

mal. In 2000, Kreuzer *et al.* published a paper summarizing their 25-year long surgical experience with the Rastelli procedure (1973-1998, 101 patients). After 20 years, 52% of the patients survived and did not require a heart transplant [12]. This percentage was greatly improved thanks to the further advancement of operative technique, better myocardial protection, changes in surgery qualification criteria, and broader experience of cardiac surgeons. In 2011, a publication by American authors appeared concerning patients operated on between 1988 and 2008. Their 20-year survival rate amounted to 93%. The risk factors for death or heart transplant included the performance of the Rastelli procedure before 1998 and the presence of extracardiac anomalies. 40% of the patients required reoperation due to RV-PA conduit stenosis. On the basis of the conducted data analysis, it was established that in 59% of cases there was no need to replace the conduit after 20 years. The risk factors for conduit replacement included young age of the patients and operation before 1988 [13].

The REV procedure

In 1982, Lecompte *et al.* proposed a technique for right ventricular outflow tract reconstruction using a patch instead of a valve conduit (REV – Réparation à l'Étage Ventriculaire) as an alternative for patients who were candidates for the Rastelli procedure [14]. The key part of this method is the excision of the conal septum, which widens the VSD and prevents LVOTO. This technique also utilizes a patch which directs blood flow from the left ventricle to the aorta. The right ventricle is directly connected to the pulmonary trunk with a unicuspid valve or without it. This method is associated with higher early mortality in comparison to the Rastelli procedure. The patients also require monitoring due to right ventricular outflow tract obstruction (RVOTO). The year of 2011 saw the publication of a work originating from Rastelli's homeland (written at the Bambino Gesù Children's Hospital in Rome). The authors (including Yves Lecompte) analyzed long-term results of 205 patients treated with the REV method between 1980 and 2003. 25-year survival was 85%; 45% of patients did not require any surgical procedure during that time. The most common reason for reoperation was LVOTO. In 77% of patients, no arrhythmias were found; 87% were in NYHA class I. In their conclusions, the authors stated that the presented surgical outcomes with the REV method were more favorable than operations based on the Rastelli procedure, both in terms of survival rate and the necessity of reoperation because of left or right ventricular outflow tract obstruction [15]. The REV method has not gained wide popularity so far, while the Rastelli procedure still remains the preferred option in the majority of cases.

The Nikaidoh procedure

In 1984, Nikaidoh presented another concept of surgical treatment for d-TGA with VSD and LVOTO: aortic translocation with reconstruction of the outflow tracts of both ventricles [16]. This method, which includes elements of

the Ross, Jatene, and Konno procedures, consists in separating the aorta from the right ventricle, removing left ventricular outflow tract obstructions by intersecting the conal septum and excising the pulmonary valve, reconstructing the left ventricular outflow tract leading to the translocated aorta with a patch appropriately sutured onto the ventricular septal defect, and reconstructing the right ventricular outflow tract using a pericardial patch. Modifications of this method include translocation of coronary vessels during aortic translocation (to prevent ischemia), the Lecompte maneuver, and reconstruction of the right ventricular outflow tract with a pulmonary homograft or the establishment of a direct connection of the right ventricle with the pulmonary artery [17, 18].

The Nikaidoh procedure – the performance of which is a technical challenge – is mainly recommended in the cases of atrioventricular canal type VSD (distant from LVOT) or restrictive VSDs, hypoplastic right ventricle, or straddling atrioventricular valve (*the valve “straddles” the interventricular septum*). In such situations, it may sometimes be the only option for biventricular correction [17-19]. It is believed that abnormal distribution of the coronary arteries constitutes a contraindication for the Nikaidoh procedure, but, in 2011, the journal *Cardiology in the Young* published a description of a successful operation with this technique performed in a patient whose anterior interventricular branch of the left coronary artery crossed the right ventricular outflow tract [20].

In 2010, a multicenter paper was published comparing the results of various methods used for the treatment of TGA with VSD and LVOTO [19]. The analysis included 146 patients undergoing different surgical procedures in the years 1980-2002 in eight European centers. In the introduction, the authors stated that the discussion concerning the surgical treatment of TGA with VSD and LVOTO is mainly focused on the comparison of outcomes between the Rastelli, Nikaidoh, and REV methods, and that there has never been published any paper that demonstrates the universal superiority of one of these procedures over the others. 82 patients underwent the Rastelli procedure (56.2%), the Jatene method (arterial switch) was performed in 24 patients (16.4%), REV in 7 patients (4.8%), atrial switch in 5 patients (3.4%), and the Nikaidoh procedure in 4 patients (2.7%). The early mortality rate was higher in the case of atrial or arterial switch with the closure of VSD and the relief of LVOTO. The late mortality rate was lower in the group of patients operated on using the REV technique and its modification with the Metras method. Reoperation due to RVOTO was most frequent in the case of patients undergoing the Rastelli procedure (37.7% of patients), followed by the REV method (14.3%) and arterial switch (13%). The highest percentage of patients not requiring reoperation or percutaneous intervention was found in the group undergoing the Nikaidoh procedure, followed by REV and the Metras modification. Reinterventions were more common in the case of LVOTO than of RVOTO. The mortality risk factors were the age of the operated children, VSD “non co-

mitted” to the aorta or pulmonary artery, and the duration of the procedure (extracorporeal circulation). The type of the procedure was not a risk factor for mortality. The age of the patients, year of the operation, and type of the procedure constituted significant prognostic factors with regard to the necessity of reoperation or percutaneous intervention. A higher risk of reoperation was found in patients undergoing the Rastelli procedure. In the REV method and its modification, conduit implantation is avoided and, thanks to the excision of the conal septum, a simple connection between the left ventricle and the aorta is achieved, which, unfortunately, does not exclude the possibility of future reoperation due to LVOTO. In the cited work the number of patients undergoing the Nikaidoh procedure was too small (4 children), and the follow-up period was too short to draw any far-reaching conclusions. In conclusion, the authors of the aforementioned multicenter publication claim that their analysis confirmed the previously reported less-than-optimal results of the Rastelli procedure, which – in comparison to the REV method and its Metras modification – requires reintervention more often, especially due to RVOTO [19].

Rastelli, Nikaidoh, or REV?

The surgical treatment of TGA with VSD and LVOTO still remains a challenge because of the necessity of reconstructing the outflow tracts of both ventricles. There seems to be a tendency to perform certain procedures in certain countries. The REV technique of Yves Lecompte and its modification with the method by Dominique Metras was initially widespread only in francophone countries, while the Rastelli procedure was popularized all around the world. The youngest of the used methods – the Nikaidoh procedure – comes from Japan, but it originated in France (a publication by Jean-Pierre Bex and Yves Lecompte in 1979). It is believed that the anatomical effect ensured by the Nikaidoh method is “closer to the norm” and provides a better location of the left and right outflow tracts and a lower risk of pressure exerted by the sternum on the right ventricular outflow tract in comparison to the Rastelli procedure (non-anatomical branching of the conduit from the right ventricular outflow tract in the Rastelli procedure). Abnormalities of coronary arteries constitute a contraindication for the Nikaidoh method. A posterior, intramural location of a coronary artery poses difficulties in translocating the aorta to the back, towards the pulmonary annulus. This type of anatomy may also prevent the transplantation of coronary arteries in conjunction with aortic translocation. In such situations, the Rastelli procedure seems to be a better option [19-22].

The Cardiac Surgery Clinic at the Children's Memorial Health Institute in Warsaw is the location of the EACTS (European Association for Cardio-Thoracic Surgery) Congenital Database. Moreover, it also runs the National Register of Cardiac Surgery. According to the EACTS data, in the five best centers in the world, the mortality rate for operations performed using the Rastelli procedure is 6.6% (15/225

patients), while for operations using the REV procedure it is 4.35% (5/115 patients). The Nikaidoh procedure was included in the EACTS database under the “other” category (together with, for example, the Kawashima procedure); the mortality rate of this group is 10.53% (6/58 patients).

The Rastelli, Nikaidoh, and REV procedures constitute three complementary options of surgical treatment. Each of these methods has its advantages and drawbacks, limitations and specific indications. There are patients who are better served by the Nikaidoh procedure than the Rastelli procedure. In the case of restrictive VSD and the impossibility of sufficient excision of the conal septum, the Nikaidoh procedure is a better option. The REV technique has similar contraindications as the Rastelli method. Each of these three options for surgical treatment has found a place in cardiac surgery, and the choice of an optimal method may sometimes be controversial with regard to mortality rates and long-term outcomes. However, for the vast majority of centers in the world, the Rastelli procedure still remains the method of choice [19-22].

Epilogue

Between 1968 and 1969, Giancarlo Rastelli received two gold medals of the American Medical Association – for his research on AVSD and for creating an innovative surgical technique for the treatment of persistent truncus arteriosus and transposition of the great arteries. He did not want to renounce his Italian citizenship, but after spending 7 years in the United States, in accordance with US law, he had to accept American citizenship. The solution turned out to be a special decree issued by President Nixon especially for him. In 1959, before his trip to the USA, Rastelli met Anna Anghileri, who was 19 at the time, during a skiing camp in the Alps. He wrote letters to her from America almost every day. Five years later, he took a flight to Europe, to Italy, and one day after arrival he married Anna (August 12, 1964). Soon after, Rastelli diagnosed himself with Hodgkin's lymphoma. He was too intelligent not to know what it meant... [1-4].

Giancarlo Rastelli loved the mountains and classical music – especially Verdi, Vivaldi, and Purcell. He had the appearance of a romantic lead and was liked by everyone. He was not able to help many children with complex heart defects. Still, he claimed that a doctor can always give these children something from himself, even if it is just several minutes of daily attention, a visit to their room, a smile, or a handshake. He was very devoted to his patients – especially Italian children pilgrimaging with their parents from Europe in the hope that American doctors would fix their incorrectly built hearts. Dr Rastelli cared about these children, invited them home, fed them, and helped them collect the funds necessary for the operation. He had a poster in his office with *l'amore vince* (love conquers) written in Italian, on which he collected the signatures of his Italian patients [2, 4, 23].

When he was losing his strength because of the progression of his disease, his sister – Rosangela – advised him to slow down and rest. He did not want to, he refused. He wrote that not working meant not living for him. John Kirk-



Phot. Giancarlo Rastelli at the age of 27. This photograph was included by Rastelli in his application documents for the NATO scholarship at the Mayo Clinic in 1960. The photograph was provided to the authors by Rastelli's daughter, Dr Antonella Rastelli, from the Washington University School of Medicine in Saint Louis, USA

lin, Rastelli's mentor and teacher, said the following several days after his death: *Perhaps the most remarkable aspect of this man's life was his reaction to his fatal illness. About five years ago, he walked into my office and said that he had Hodgkin's disease. He told me this with about the same display of emotion that he would have used should he have said that our densitometer was not working properly. Somehow an unspoken agreement developed between us that neither of us would speak of this illness unless there was urgent need to do so. About a year and a half later, another recurrence developed and he told me of this in the same simple words. The serenity and confidence with which he faced life and death is the greatest of the many things that he taught me.* [1, 2, 23, 24].

Giancarlo Rastelli died in a Rochester hospital after a five-year struggle with the disease, 4 months before his 37th birthday. He orphaned his 4-year-old daughter, Antonella Luisa (she is currently a doctor and works in St Louis). His body was brought to Italy and buried in the chapel of the University of Parma. 35 years later – on September 30, 2005 – his beatification process began in the Vatican. The life of Rastelli was considered an example and inspiration for young Catholics, medical students, and novice physicians.

Those who know his biography believe that his attitude towards patients, life, and being a doctor is an example to follow. The most important issue in the beatification process is to prove the extraordinary nature of the life of the person who died in the aura of sanctity. It is related to the so-called “heroism of virtues” (*heroicas virtutum*). The petitioner of Rastelli's beatification is the Diocese of Parma and the Association of Catholic Doctors. The task will be to prove, among other things, the heroism of Giancarlo Rastelli's virtues. The beatification process is led by the Congregation for the Causes of Saints (protocol no. 2678). Time will show whether the cardiac surgeon will become a saint... [1-4, 23, 24].

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