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Italian survey on cardiac surgery for adults with congenital heart disease: which surgery, where and by whom?

Alessandro Giamberti^{a,*}, Massimo Chessa^b, Carmelina Chiarello^a, Adriano Cipriani^c, Adriano Carotti^d, Lorenzo Galletti^e, Gaetano Gargiulo^f, Stefano Maria Marianeschi^g, Carlo Pace Napoleone^h, Massimo Padalinoⁱ, Gianluigi Perra^j and Giovanni Battista Luciani^k, on behalf of the Congenital Domain of the Italian Society of Cardiac Surgery

^a Department of Congenital Cardiac Surgery, IRCCS Policlinico San Donato, San Donato M.se, Italy

^b Pediatric and Adult Congenital Heart Centre, IRCCS Policlinico San Donato, San Donato M.se, Italy

^c Centro per la Cura delle Cardiopatie Congenite dell'Adulto, Istituto Clinico Ligure di Alta Specialità (ICLAS), Rapallo, Italy

^d Department of Pediatric Cardiac Surgery, Bambino Gesù Childrens Hospital IRCCS, Roma, Italy

^e Pediatric Cardiology and Cardiac Surgery Unit, Cardiovascular Department, Papa Giovanni XXIII Hospital, Bergamo, Italy

^f Pediatric and Grown-up Congenital Cardiac Surgery, University of Bologna, S.Orsola-Malpighi Hospital, Bologna, Italy

^g Pediatric Cardiac Surgery Unit, Grande Ospedale Metropolitano Niguarda, Milano, Italy

^h Pediatric Cardiac Surgery, Regina Margherita Children's Hospital, Torino, Italy

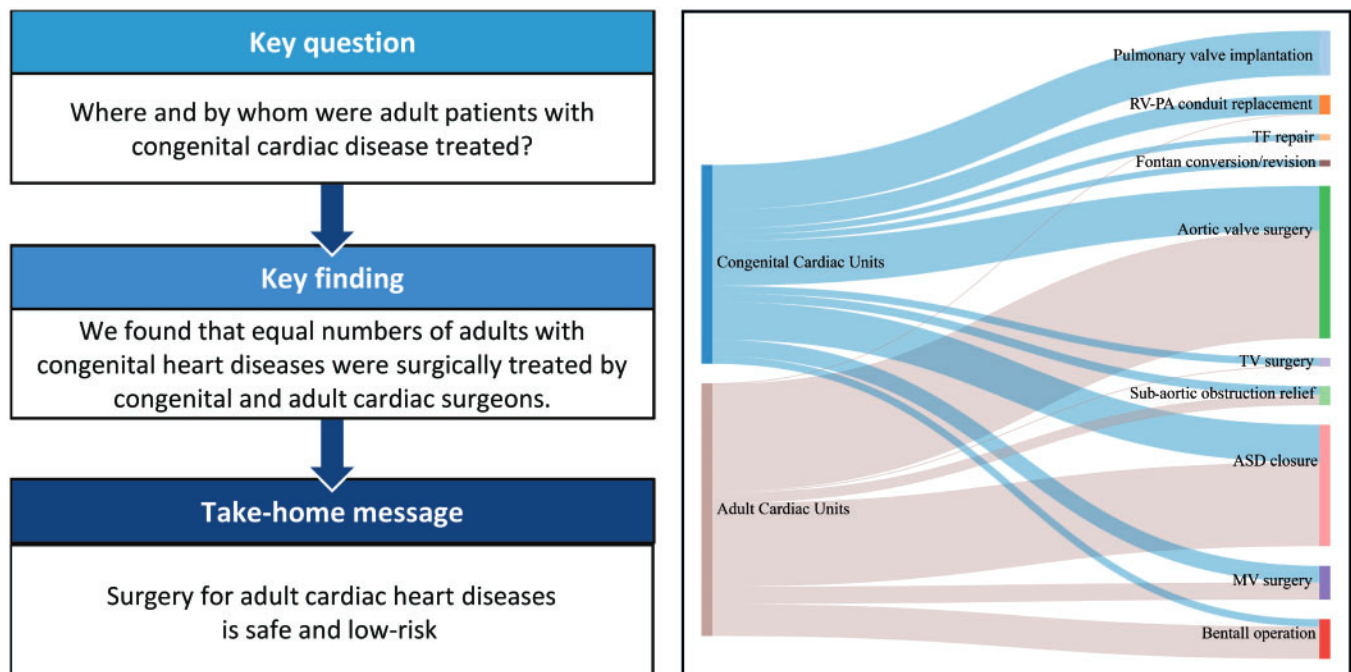
ⁱ Pediatric and Congenital Cardiac Surgical Unit, Department of Cardiac, Thoracic and Vascular Sciences, Padova University Hospital, Padova, Italy

^j Department of Cardiac Surgery, Policlinico Gemelli Hospital, Roma, Italy

^k Department of Surgical Sciences, Dentistry, Gynecology and Pediatrics, Division of Cardiac Surgery, University of Verona, Verona, Italy

* Corresponding author. Department of Congenital Cardiac Surgery, IRCCS Policlinico San Donato, Via Morandi 30, 20097 San Donato M.se, Italy. Tel. +39-02-52774677; fax: +39-02-25774459; e-mail: alegia@hotmail.com (A. Giamberti).

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[†]Chairman of the Congenital domain of the Italian Society of Cardiac Surgery (SICCH).

Abstract

OBJECTIVES: The population of ageing adults with congenital heart disease (ACHD) is increasing; surgery in these patients presents major difficulties in management. A great debate has developed about whether these patients should be cared for at an adult or paediatric hospital and by an acquired or congenital cardiac surgeon. We analysed data of the surgical treatment of ACHD from the Italian cardiac surgery centres in 2016, focusing on the type of surgery performed, where these patients were operated on and by whom.

METHODS: Ninety-two Italian cardiac surgery centres were contacted and 70 centres participated in this study. We collected data on the types of cardiac operations performed in congenital heart defect patients older than 18 years. In 2016, a total of 913 patients with ACHD were operated on: 440 by congenital cardiac surgeons (group I) in centres with paediatric and adult cardiac surgery units, and 473 by adult cardiac surgeons (group II) in centres with exclusively adult cardiac surgery units.

RESULTS: Pathologies of the right ventricular outflow tract were the most frequent diseases treated in group I and pathologies of the left ventricular outflow tract in group II. Group I included more complex and heterogeneous cases than group II. Surgery for ACHD represented 12% of the activity of congenital cardiac surgeons and only 1% of the activity of adult cardiac surgeons.

CONCLUSIONS: In Italy, ACHD patients are operated on both by congenital and adult cardiac surgeons. Congenital cardiac surgeons working in centres with both paediatric and adult cardiac surgery are more involved with ACHD patients and with more complex cases.

Keywords: Adult with congenital heart disease • Cardiac surgery • Grown-up congenital heart disease

INTRODUCTION

Thanks to improved surgical and interventional cardiac procedures in the last forty years, over 90% of the children born with a congenital heart defect (CHD) can be expected today to reach adulthood.

Adults with congenital heart disease (ACHD) are a growing population [1] and recent data [2, 3] estimate that, in Europe, we are currently faced with an ACHD population of 2.3 million which significantly outnumbers the paediatric CHD population of 1.9 million patients.

Cardiac operations in these patients present major difficulties in management and techniques. Many of these patients have undergone complete repair in neonatal or paediatric life, and need to be reoperated for residual lesions, prosthetic materials degeneration or recurrent haemodynamic sequelae. Another group includes patients with non-repaired malformation reaching adulthood experiencing their natural history or after palliative procedures. Finally, a third group includes patients who need to be reoperated for acquired heart disease (AHD) unrelated to the native malformation, such as coronary artery disease. The latter group of patients is the smallest, but is expected to increase with the growing number of patients who reach the age of 50 or older [4, 5].

An ongoing debate in the literature is whether these patients should be cared for at an adult or paediatric hospital and by an adult or paediatric cardiac surgeon, with the relative advantages and disadvantages of each system [6–9].

The aim of this study, carried out on behalf of the Congenital Domain of the Italian Society of Cardiac Surgery, was to analyse data from the Italian cardiac surgery centres in order to have a picture of the Italian scenario in this field.

MATERIALS AND METHODS

We conducted a descriptive and retrospective survey of ACHD patients operated on in Italy in the year 2016. Ninety-two centres were identified through the Italian Society for Cardiac Surgery

website: 16 cardiac surgery centres dedicated to CHD and 76 adult cardiac surgery centres dedicated to AHDs. Currently, there is no official database for cardiac surgery in Italy and the only data available from the Ministry of Health were the number of patients operated on in individual centres. In this context, the only possible method of investigation was sending an email to the respective head of the cardiac surgery units.

Inclusion criteria were cardiac operation performed in CHD patients older than 18 years. Adult congenital cardiac surgical procedures were defined as those performed for a cardiac defect present from birth and that it was the main indication for the surgery. Bicuspid aortic valve disease patients were included up to a maximum age of 35 years, in accordance with the National Congenital Heart Disease Audit Website in the UK [10]. Patients with patent foramen ovale were excluded. Patients with acquired cardiac disease unrelated to the congenital defect were excluded.

For each patient, the principal diagnosis was considered, and the CHD was classified in terms of complexity according to the Bethesda score [11].

We identified 2 groups of patients: patients operated on by congenital cardiac surgeons (group I) and patients operated on by adult cardiac surgeons (group II). The surgeon was designated as a congenital or adult cardiac surgeon by the nature of one's own surgical practice.

This research was conducted to better understand where ACHD patients in Italy had been operated on, and by whom, the type of surgery performed and the outcomes.

Statistical analysis

Data collection was performed through a survey search realized by written inquiry. Descriptive statistics for categorical variables are reported as frequency and percentage, whereas continuous variables are reported as mean with standard deviations. Categorical variables in the 2 groups were compared using the χ^2 test (SPSS, Inc., Chicago, IL, USA). Statistical difference was considered significant at a level of P -value <0.05.

RESULTS

Group I: patients operated on by congenital cardiac surgeons

The cohort of study included 913 ACHD patients, 440 of whom (group I) were treated in the 16 Italian cardiac surgery units dedicated to CHD. Among these, 3 units operate either in a children's hospital or paediatric service, 2 adult units host ACHD procedures with congenital heart surgeons coming in from children's hospitals, while 11 units operate in institutions equipped with both adult and congenital cardiac surgery services.

In group I, out of 3575 patients with CHD operated on in 2016 in these 16 centres, ACHD procedures accounted for 12.3% of overall activity. The number of ACHD surgical procedures performed in the different centres ranged from 7 to 80 operations per year (mean 28/year). Surgical activity on ACHD patients ranged from 6% to 30% of the overall activity of each individual centre. Five centres operated on more than 30 ACHD patients in 2016 and 2 centres operated on more than 50 cases (Fig. 1).

Data on different diagnoses and type of operations performed were collected from all the centres and are summarized in Tables 1 and 2. Pathology of the right ventricular outflow tract was the most frequent (29%) including 78 surgical pulmonary valve implantations, 33 conduit replacements and 11 tetralogy of Fallot repairs.

Pathology of the left ventricular outflow tract was present in 24% of cases including 78 interventions for aortic valve disease, 15 subaortic obstruction relief and 13 Bentall procedures. Atrial septal defect (ASD, including secundum, primum and sinus venosus) repair was performed in 68 patients (15%). Less frequently, mitral valve repair/replacement was performed in 6.5% of patients, partial anomalous pulmonary venous drainage repair in 5%, tricuspid valve repair/replacement in 3%, Fontan conversion/revision in 2.7%. Coronary artery bypass grafting (CABG) was performed in 6 cases ($6/440 = 1.5\%$), and was associated with ASD closure in 4, pulmonary valve implantation in 1 and partial anomalous pulmonary venous drainage repair in 1 (Table 1).

According to the type of operation performed, 238 patients (54%) had a reoperation after previous repair, while 190 patients (43%) had primary repair.

Eight patients (2%) had completion of repair after previous palliation, and 4 patients (1%) had a second palliation after first-stage palliation. The types of surgical procedures performed are shown in Table 2.

Hospital mortality was 1.5% ($n = 6$ patients). One patient died after extra-cardiac Fontan conversion due to low cardiac output syndrome. Two patients previously treated with extra-cardiac Fontan conversion died after ventricular septal defect (VSD) enlargement for management of subaortic obstruction. Finally, 3 patients died of right ventricular failure after right ventricular outflow tract obstruction (RVOTO) relief and pulmonary valve implantation.

Group II: patients operated on by adult cardiac surgeons

We invited 76 Italian adult cardiac surgery centres dedicated to ACHD patients with cardiac surgeons not operating on paediatric congenital cases to collaborate for this study. Fifty-four centres ($54/76 = 70\%$) participated and contributed to the data collection. The majority (17 of 22 centres) of non-participating centres represented small programmes.

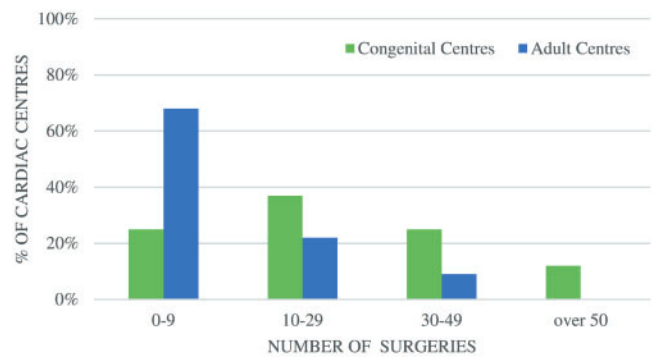


Figure 1: Comparative data of surgical activity in both groups.

Table 1: Group I: most common cardiac diagnoses

	n (%)
RVOT pathologies	129 (29)
Pulmonary valve regurgitation	78
Conduit obstruction	33
Tetralogy of Fallot	11
Pulmonary valve stenosis	7
LVOT pathologies	106 (24)
Aortic valve disease	91
Subaortic obstruction	15
ASD	63 (15)
Ostium secundum	41
Sinus venosus	17
ASD redo	3 (0.7)
Scimitar syndrome	2 (0.4)
MV disease	29 (6.5)
Partial AV canal (ostium primum)	14 (3.2)
Fontan failure	12 (2.7)
TV disease	13 (3)
Miscellaneous	69 (15.7)

ASD: atrial septal defect; AV: atrioventricular; LVOT: left ventricular outflow tract; MV: mitral valve; RVOT: right ventricular outflow tract; TV: tricuspid valve.

In 2016, a total of 31 078 adult cardiac patients were operated on in these 54 centres: 473 of them (1.5%) were ACHD.

The number of ACHD operated on in each centre ranged from 0 (11 centres) to 48 (mean 9/year). Eight centres operated on more than 12 cases in 2016, 1 centre more than 24 cases and 4 centres more than 36 cases.

Data on different pathologies and surgery performed are summarized in Tables 3 and 4.

Pathology of the left ventricular outflow tract was the most frequent (55%) including 206 procedures on the aortic valve, 56 Bentall procedures and 18 subaortic obstruction repairs. ASD closure was the most frequent procedure (146 cases, 31%) followed by surgery for congenital mitral valve abnormalities (6%). Three hundred and seventy-six patients had surgery as the first operation (79%) and 97 patients (21%) had a reoperation after a repair. CABG was performed in 19 cases (4%).

Of the 266 heart transplants performed in Italy in 2016 (data available from the Ministry of Health website), only 4 (1.5%) were performed on ACHD patients.

Similar to ACHD operations performed by congenital cardiac surgeons, cardiac valve procedures in general represented more than half of the operations in ACHD patients performed by adult cardiac surgeons. ASD repair represented 1/3 of all operations in

Table 2: Group I: most common cardiac surgical procedures performed

	n (%)
Pulmonary valve implantation	78 (18)
RV-PA conduit replacement	33 (7.5)
ToF repair	11 (2.5)
Aortic valve surgery	78 (18)
Subaortic obstruction relief	15 (3)
Bentall operation	13 (3)
ASD closure	66 (15)
Congenital MV surgery	29 (6.5)
Congenital TV surgery	13 (3)
Fontan conversion/revision	12 (2.7)
Partial AV canal repair	14 (3.2)

ASD: atrial septal defect; AV: atrioventricular; MV: mitral valve; RV-PA: right ventricle-pulmonary artery; ToF: Tetralogy of Fallot; TV: tricuspid valve.

Table 3: Group II: most common cardiac diagnoses

	n (%)
RVOT pathologies	1 (0.2)
LVOT pathologies	262 (55)
Aortic valve disease	244
Subaortic obstruction	18
ASD	146 (31)
Congenital MV disease	30 (6.3)
Congenital TV disease	2 (0.5)
Miscellaneous	32 (6)

ASD: atrial septal defect; LVOT: left ventricular outflow tract; MV: mitral valve; RVOT: right ventricular outflow tract; TV: tricuspid valve.

adult cardiac units. Hospital mortality was 0.8%. Comparative data between groups I and II are shown in Table 5.

In order to compare in both groups the patient distribution in terms of complexity of cardiac disease, we used the Bethesda classification categorizing the congenital heart diseases into 3 classes: mild, moderate and severe [11]. As reported in Fig. 2, only group I included patients with a severe index of complexity, while the majority of ACHD patients in the group II are patients with a simple heart defect, demonstrating that, when the complexity of the cardiac disease increases, the patients with moderate or severe defects were predominantly treated in cardiac surgery units by congenital surgeons.

DISCUSSION

Early diagnosis, advances in surgical and interventional technique and medical therapy have increased overall survival to adulthood of new-borns affected by moderate-to-complex CHD. The ACHD population is constantly growing and ageing with a progressive increase of patients older than 50 years [4, 5, 12, 13].

The natural ageing of ACHD patients determines an enhanced incidence of the classical age-related diseases with an important increasing number of hospital admissions. Many of these patients need to be operated on or reoperated for their CHD, or for AHD unrelated to the native malformation such as CABG [14].

There is a consensus that ACHD patients, particularly those with moderate to highly complex CHD, can be difficult to

Table 4: Group II: most common cardiac surgical procedures performed

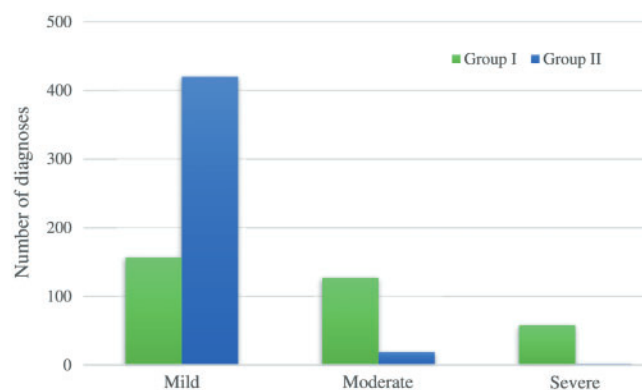
	n (%)
RV-PA conduit replacement	1 (0.2)
Aortic valve surgery	188 (40)
Subaortic obstruction relief	18 (4)
Bentall operation	56 (12)
ASD closure	146 (31)
Congenital MV surgery	30 (30)
Congenital TV surgery	2 (0.5)
Miscellaneous	32 (6)

ASD: atrial septal defect; MV: mitral valve; RV-PA: right ventricle-pulmonary artery; TV: tricuspid valve.

Table 5: Comparative data between groups I and II

	Group I	Group II	P-value
Number of patients	440	473	
Percentage of total activity	12.3	1.5	<0.001
Patients/centre (mean)	28 (SD: 22.6)	9 (SD: 12.2)	
Diagnoses (%)			
RVOT	29	0.2	<0.001
LVOT	24	55	<0.001
ASD	15	31	<0.001
MV	6.5	6.3	0.87
TV	3	0.4	0.002
Reoperation (%)	54	21	<0.001
CABG (%)	1.5	4	0.021
Mortality (%)	1.5	0.8	0.45

ASD: atrial septal defect; CABG: coronary artery bypass grafting; LVOT: left ventricular outflow tract; MV: mitral valve; RVOT: right ventricular outflow tract; SD: standard deviation; TV: tricuspid valve.

**Figure 2:** Complexity degree of adults with congenital heart disease treated in both groups using the Bethesda classification.

manage and that these patients have very special needs. This is a timely topic of increasing clinical interest suggesting that complex ACHD patients should be treated and managed in a few experienced and specialized 'grown-up congenital heart' (GUCH) units where these patients are frequently treated, concentrating resources, patients, funding and professional experience, as recommended by the American Heart Association and the American College of Cardiology [14].

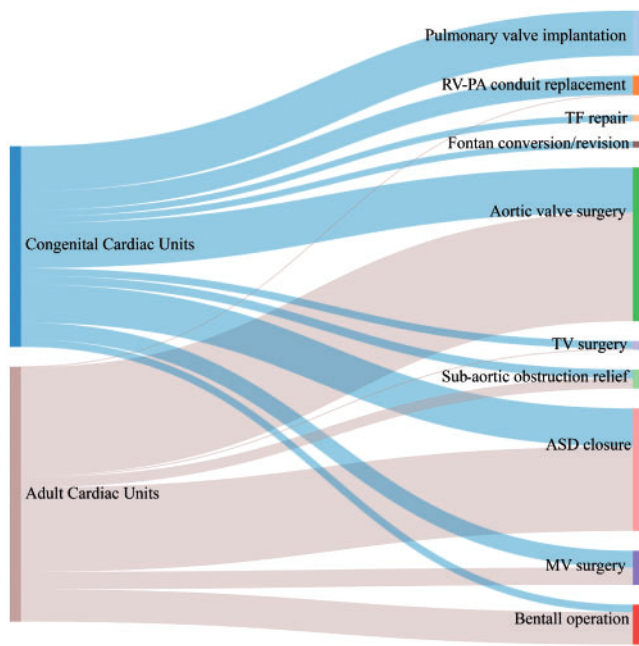


Figure 3: Sankey plot showing the adults with congenital heart disease surgical activity for both groups. ASD: atrial septal defect; MV: mitral valve; RV-PA: right ventricle-pulmonary artery; TF: tetralogy of fallot; TV: tricuspid valve.

The European Society of Cardiology (ESC) through the Working Group on GUCH has recently published practice guidelines for the management of GUCH patients [15, 16] and recommendations for the organization of care for ACHD in Europe [16].

Few data are available about surgery for ACHD and the ideal model of the care for the surgical patients does not exist at the present time.

In 2003, the Congenital Heart Disease Committee of the European Association of Cardio-Thoracic Surgery published the indications and the suggestion for the 'optimal structure of a congenital heart surgery department in Europe' [17]. With regard to surgical care for ACHD, the authors observe that many congenital heart surgery units are today developed in children's hospitals where the treatment of adolescent or adult patients is difficult and they concluded that larger centres with facilities for congenital and for adult cardiac surgery are in a good position to treat these patients.

An overview of the management of ACHD in the United States was presented at the Society of Thoracic Surgeons Association meeting in 2007 [7]. They analysed 40 000 adult congenital cases and, very surprisingly, the authors discovered that congenital surgeries performed only 5% of the adult cases and that 95% of ACHD patients were operated on by non-congenital heart surgeons (adult cardiac surgeons). The majority of the procedures were simple lesions, but more complex lesions were also included. At the same time, it was clearly demonstrated that early mortality was significantly lower if the operation was performed by a congenital cardiac surgeon, and that an independent risk factor for early mortality included the operation being performed by a non-congenital heart surgeon. Furthermore, the study demonstrated a decrease in hospital length of stay and a lower hospital charges when GUCH surgery was performed by a congenital heart surgeon [7].

The Canadian Cardiovascular Society guidelines for the management of ACHD [18] and the American College of Cardiology/American Heart Association Guidelines for the management of ACHD [14] indicate that for all congenital diagnosis, the

operations should be performed by surgeons with training and expertise in congenital heart disease.

The optimal environment to operate on these patients (adult or children's hospital) is another matter of debate [8, 9, 19, 20]. Kogon *et al.* [8] demonstrated that adult patients with CHD should have their care at an adult institution with surgery being performed by a congenital cardiac surgeon. Another study by Karamlou *et al.* [9] confirmed this recommendation. The constantly increasing number of patients needing complex surgery for ACHD and the high incidence of reoperation favour having the surgery performed by congenital cardiac surgeons. At the same time, the constant increase in age of these patients, frequently older than 50, and the possible presence of concomitant AHD and/or comorbidities (endocrine, rheumatological, hepatological, neurological and renal) are valid arguments supporting the thesis to prefer an adult environment for the care of these patients. Finally, we should take into consideration that patients in their 30s, 40s, 50s and older wanting to be operated on in an adult centre, and the fact that paediatric nurses at children's hospitals prefer to care for children rather than adults. As suggested by Vouhé [20], the best solution should be to create specialized centres where adult and paediatric cardiac surgery facilities are developed and where expert congenital surgeons (with a large paediatric cardiac surgery practice) perform a critical volume of surgery for adult patients with occasional collaboration of adult cardiac surgeons for specific procedures when corresponding training is missing. The observation that more than half of the operations required in ACHD involve repair or replacement of cardiac valves (aortic, mitral, pulmonary, tricuspid and trunclal), as evident from the present survey and from prior studies [21], begs the question of the optimal training of the next generation of congenital cardiac surgeons. Clearly, additional training in 'structural heart disease', starting with valve disease, including open and catheter-based procedures must be offered to surgeons, who must face the challenges of CHD beyond childhood and into adolescence and adulthood [22].

In this study, we sought to analyse the current scenario in Italy. The number of patients operated on in groups I and II is practically the same, but it is clear that surgery for ACHD had a significantly different impact for paediatric cardiac surgeons (12.3% out of 3575 patients) than on adult cardiac surgeons (1.5% out of 31 078 patients, $P < 0.0001$).

Based on our data, we can assert that paediatric cardiac surgeons operated more frequently right-sided heart lesions, reoperations, and more complex and heterogeneous lesions while adult cardiac surgeons were more involved in the primary treatment of aortic valve disease and ASD patients. Figure 3 summarizes the trend of all our data related to both groups. Incidentally, this observation on differing indications and procedures in ACHD patients operated on in 'spoke' units (adult cardiac units) when compared to 'hub' units (congenital cardiac units) is in line with recommendations of the AHA/JACC [14].

Finally, our study demonstrated that surgical treatment of these patients is safe and low risk regardless of whether these patients are operated on by paediatric or adult cardiac surgeons. The results presented in this study suggest further improvement of previously published data [23, 24], confirming that the greater experience acquired through the years has contributed to decrease operative mortality.

Limitations

There are several limitations in this study. Firstly, this study is retrospective in nature and subject to limitations inherent to

observational investigations and to methodology to collect all data. Secondly, we analysed a very short period of time and, consequently, this study is just a picture of the Italian scenario related only to 2016. Thirdly, the study is limited to the 76% of the Italian centres performing cardiac surgery. In particular, while all Congenital Cardiac Units provided complete data, only 70% of Adult Cardiac Units provided data. This limitation is related to the fact that currently there is no official database for cardiac surgery in Italy. The majority of non-participating centres represented small programmes; therefore, we believe that our sample is representative of all centres in Italy and that the study findings can be generalized.

CONCLUSIONS

ACHD patients are operated on today in Italy both by congenital and adult cardiac surgeons. Congenital cardiac surgeons working in centres with both paediatric and adult cardiac surgery units seem to be more involved in the surgical treatment of ACHD patients and operate more frequently on right-sided heart lesions, reoperations and more complex cases while adult cardiac surgeons are primarily dedicated to ACHD patients with aortic valve disease and ASDs. Although this is only one year's collection, our data analysis shows that surgical treatment for ACHD patients is safe and at low risk, and the results obtained demonstrated a marked improvement compared to previously published data [20, 23].

SUPPLEMENTARY MATERIAL

Supplementary material is available at *ICVTS* online.

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Conflict of interest: none declared.

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