Anesthetic Management of Congenital Heart Disease "Right to Left Shunt" In Adults Undergoing Non Cardiac Surgery

Essay

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Engy Wagdy Wissa Marcos

Contents

List of Abbreviations

ABGs	:	Arterial blood gases
ACC/AHA	:	American College of Cardiology/American Heart Association
ACHD	:	Adult (with) congenital heart disease/s
ACOG	:	American College of Obstetrics and Gynecology
AHA	:	American Heart Association
A-P	:	Aorto-pulmonary
ASA	:	American Society of Anesthesiologists
ASD	:	Atrial septal defect
AV	:	Atrioventricular
AVSD	:	Atrioventricular septal defect
BP	:	Blood pressure
B-T shunt	:	Blalock-Taussig shunt
CCL	:	Cardiac catheterization laboratory
CHD/s	:	Congenital heart disease/s
CHF	:	Congestive heart failure
CONCOR programme	:	Dutch National Registry and DNA-Bank of Patients with Congenital Heart Disease in the Netherlands, named CONCOR (CONgenital CORvitia)
COP	:	Cardiac output
CRP	:	C- reactive protein
C-section	:	Caesarean section
CT	:	Computed tomography
CVP	:	Central venous pressure
2 D-echo	:	Two dimensions-echocardiography
d-TGA	:	Dextro-transposition of great arteries
ECG	:	Electrocardiography
ECHO	:	Echocardiography
ENT	:	Ear, Nose and throat
ESC	:	European Society of Cardiology
ETT	:	Endo-tracheal tube

List of Abbreviations (Cont.)

FiO_2	:	Fractional inspired oxygen concentration
FRC	:	Functional residual capacity
Hct	:	Hematocrit
HIT	:	Heparin induced thrombocytopenia
HR	:	Heart rate
HTN	:	Hypertension
IART	:	Intra-atrial re-entrant tachycardia
ICD/s	:	Implantable cardioverter-defibrillator/s
ICU	:	Intensive care unit
IE	:	Infective endocarditis
IM	:	Intra-muscular
iNO	:	Inhalational nitric oxide
INR	:	International Normalized Ratio
IPPV	:	Intermittent positive-pressure ventilation
IV	:	Intra-venous
IVC	:	Inferior vena cava
L	:	Left /Levo
LA	:	Left atrium /atrial
LAP	:	Left atrial pressure
LMA	:	Laryngeal mask airway
LV	:	Left ventricle
LVOT	:	Left ventricular outflow tract
MAPCAs	:	Major aorto-pulmonary collateral arteries
MPA	:	Main pulmonary artery
MRI	:	Magnetic resonance imaging
NPO	:	Nil per os = Nothing by mouth
NYHA	:	New York heart association
OR	:	Operating room
PA/IVS	:	Pulmonary atresia with intact ventricular septum
PA/s	:	Pulmonary artery/ies
PA/VSD	:	Pulmonary atresia with ventricular septal defect

List of Abbreviations (Cont.)

$PaCO_2$	:	Arterial CO_2 tension
PaO_2	:	Arterial oxygen tension
PAP	:	Pulmonary artery pressure
PAPVD	:	Partial anomalous pulmonary venous drainage
PAPVR	:	Partial anomalous pulmonary venous return
PBF	:	Pulmonary blood flow
PCWP	:	Pulmonary capillary wedge pressure
PDA	:	Patent ductus arteriosus
PEEP	:	Positive end-expiratory pressure
$P_{ET}CO_2$	:	End-tidal carbon dioxide
PFO	:	Patent foramen ovale
PHT	:	Pulmonary hypertension
PPCs	:	Postoperative pulmonary complications
PR	:	Pulmonary regurgitation
PS	:	Pulmonary stenosis
PT	:	Prothrombin time
PTT	:	Partial thromboplastin time
PVR	:	Pulmonary vascular resistance
Q_P	:	Pulmonary blood flow/perfusion = blood flow through the lungs
Q_P/Q_S	:	Pulmonary to systemic blood flow (perfusion) ratio
Q_S	:	Systemic blood flow/perfusion
RA	:	Right atrium/atrial
RAP	:	Right atrial pressure
RBBB	:	Right bundle branch block
R-L, R-to-L	:	Right-to-left
RV	:	Right ventricle/ventricular
RVOT	:	Right ventricular outflow tract
SaO_2	:	Arterial oxygen saturation
S-P	:	Systemic-to-pulmonary
SpO_2	:	Oxygen saturation
SV	:	Systemic ventricle

List of Abbreviations (Cont.)

SVC	:	Superior vena cava
SVR	:	Systemic vascular resistance
TAPVD	:	Total anomalous pulmonary venous drainage
TAPVR	:	Total Anomalous Pulmonary Venous Return
TEE	:	Trans-esophageal echocardiography
TGA	:	Transposition of the great arteries
TOF	:	Tetralogy of Fallot
vs.	:	Versus
VSD	:	Ventricular septal defect
VT	:	Ventricular tachycardia
WBC	:	White blood cell
WPW	:	Wolff-Parkinson-White syndrome

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Introduction

Congenital heart defects are the most common group of inborn defects, occurring in approximately 8 in 1,000 live births. It is estimated that approximately 30,000 children are born annually with congenital heart diseases (CHD). In the era before the development of pediatric cardiac surgery, the majority of untreated patients born with congenital heart disease die in infancy or childhood, and only 15-25% survive into adulthood. Those who survive were the patients who have a mild form of the disease that has allowed them to survive without surgical or interventional cardiac catheterization. ***(Deanfield et al., 2003)***

Advances in prenatal diagnosis, interventional cardiology, paediatric cardiac surgery, anesthesia, and critical care over the past 40 years have resulted in survival of approximately 90% of these children beyond their third decade of life. This dramatic success has created a rapidly increasing population of adult patients with congenital heart disease (CHD) so the vast majority of the patients with CHD -seen in the adult outpatient setting- are patients who have had previous early surgical definitive repair, catheter-based intervention, palliative surgery or even heart transplantation as their only option. ***(Fleisher et al., 2007)***

The spectrum of CHD ranges widely from relatively mild defects -seen in simple isolated defects- to lesions of moderate to severe complexity typically characterized by several coexistent malformations. As regards adult patients with CHD, they can broadly be divided into three categories: those who have undergone some sort of reparative operation; those who have undergone a palliative operation; and finally, there is a group of patients who have not previously undergone a corrective or palliative procedure. Survival rates in CHD are influenced by many factors, including year of birth, age at diagnosis, complexity of the pathology, and whether the

lesion(s) has been palliated or surgically corrected, taking in consideration that adult patients with structural heart disease who have undergone repair of what are considered uncomplicated lesions may be indiscernible from unaffected patients while other patients with unrepaired CHD -who were not considered to have hemodynamic compromise worthy of intervention -have incurred severe physiologic perturbations as sequelae of their unrepaired CHD and no longer candidates for repair of their structural heart lesions and this results in patients with complications who are constituting an extremely high risk group. ***(Weiss, 2004)***

Adult patients with moderate to severe CHD undergoing non-cardiac surgery should be referred and followed up in a comprehensive adult CHD centre. This was recommended by a task force of the American College of Cardiology, American Heart Association and the Canadian Cardiovascular Society. These centres include cardiologists, heart surgeons, and cardiac anaesthesiologists with specialized training and extensive experience in the field so the patient would obtain the appropriate consultation; and where there are intraoperative trans-oesophageal echocardiography availability; and the full range tests of cardiac function and perfusion; for adequate assessment, monitor and management. But unfortunately as this population grows, patients will inevitably seek urgent care in a variety of centres, including some that do not meet all of the standards set out by the American Heart Association. ***(Therrien, Gatzoulis et al., 2001)***

The most frequently encountered coexisting medical disorders -that may be a direct result of CHD-induced physiology or a result of age-related acquired derangement- should be considered as well as the anaesthetic and surgical techniques employed in these procedures and their hemodynamic consequences. Also the medications required for these unique patients need to be considered along with

their inherent side effects. Stratification into high- or low-risk surgery must be well established where it is based on whether the operation is elective or emergent and major or minor. ***(Weiss, 2004)***

Here we are especially focusing on CHD (right-to-left shunting), that include those anatomical heart defects that produce a limitation in pulmonary blood flow or result in mixing of oxygenated and deoxygenated blood. Both conditions lead to decreased blood oxygen content and cyanosis. Unlike the acyanotic forms of congenital heart disease, the majority of patients with cyanotic congenital heart disease will have had at least one and often several previous interventions before adulthood. With time, compensatory physiology will evolve and produce abnormal loading conditions and possible impairment in ventricular function. Many of the compensatory changes are imperceptible and cause little impairment in function, whereas others cause significant cardiovascular compromise. ***(Khairy et al., 2007)***

A primary goal of this essay is to care for adult with congenital heart disease (ACHD), to diminish cardiac-related morbidity and to avoid adverse perioperative events during undergoing non-cardiac surgery in the presence of uncorrected, palliated, or corrected lesions. Of utmost importance in this essay is having a basic understanding of the most common congenital cardiovascular defects as applied to the adult age group regarding the native anatomy, physiology, surgical strategies, and late outcome of the defect under consideration.

The Aim of the Essay

The ultimate goal of this essay is to provide the anesthesia care provider with an understanding of the basic underlying anatomy and associated physiology of the most common congenital heart defects -with right to left shunts- as applied to the adult age group. It also provides an overview of the long-term consequences and the preoperative and intraoperative implications of those CHD when undergoing non-cardiac surgery and the potential hemodynamic consequences of various anaesthetic and surgical techniques; in order to diminish cardiac-related morbidity and to avoid adverse perioperative events. This will assist the anesthesia care provider in formulating a pre-operative plan and approaching any hemodynamic perturbations with the appropriate interpretation and management.

Chapter One

Pathophysiology of Adult Congenital Heart Diseases with Right-to-Left Shunt

Congenital heart diseases (CHDs) are common pathologies because they occur in 0.5% to 1% of births; among them, complex malformations -which include most of CHD with right-to-left (R-to-L)-are less frequent (0.15% of births). The recent major advances made in congenital cardiac surgery have resulted in an increased number of children born with CHD who are expected to reach adulthood. This made this population growing at a rate of 5% each year. ***(Park, 2010)***

Although patients with ACHD have an overall decreased life expectancy when compared with individuals with normal hearts, yet the 15-year survival rate is considered high being 80% and 95% for complex and simple CHD, respectively; half of the patients with complex physiology are over 25 years old. Any anesthesiologist might therefore encounter one of these patients in his daily practice. His role is pivotal in their management, during non-cardiac surgery and obstetrics. With his good understanding of the pathophysiology, and his working knowledge of how to deal with these CHDs patients, he can lead the decisions of the operative team. ***(Brickner et al., 2000)***

Patients having adult congenital heart disease (ACHD) with R-to-L present to adult physicians with the lesion distribution declared in the Dutch national registry (CONCOR programme) as shown in Table (1).