Cardiac Catheterization for Congenital Heart Disease

From Fetal Life to Adulthood

Gianfranco Butera
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Editors

Second Edition

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Foreword: "Nothing Endures But Change"

This concept was stated by Heraclitus of Ephesus in his book *On Nature* in the sixth century B.C. A long time has passed from the period of this pre-Socratic philosopher; however, the same concept applies today and truthfully to the field of interventional cardiology. The only way to be alive and up-to-date is to continuously improve and change. Therefore, it is with this thought we welcome the second edition of the *Cardiac Catheterization for Congenital Heart Disease*. The second edition follows the popular "how-to" format of the previous edition and is the result of a worldwide cooperation of many international experts, who put together their knowledge and expertise.

The first edition was published in 2014 and many new transcatheter therapies have been developed in the field of interventional cardiology for CHD, including the expanded use of balloon-expandable valves for dysfunctional RVOTs, conduits, and valve-in-valve therapies in all four cardiac valves. The initial use of self-expanding valves in the RVOT to treat severe pulmonary regurgitation has continued to be developed around the globe. The increased role of advanced imaging, such as 3D Rotational Angiography (3DRA), has allowed interventional cardiologists to use fluoroscopic overlay to not only improve the precise positioning of devices such as stents and valves but also to potentially decrease radiation exposure by reducing the need of additional angiograms.

The role of integrating advanced imaging modalities in the cath lab, such as real-time MR-guided cardiac catheterizations and potential interventional procedures without the use of radiation, has been developed and promoted by centers around the world. In addition, 3D printing has become popular in both planning complex surgical and interventional procedures, and educating patients and families in their complex CHD. Finally, the editors have included a much-needed section on quality improvement of both results and care in our ongoing quest of delivering the highest form of interventional cardiology for our patients.

As Heraclitus stated "nothing endures but change," he also said "the eyes are more exact witnesses than the ears"…which is the way interventional cardiologists view improving our procedures to achieve our core value and mission of treating our patients.

Respectfully,

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Preface

"Our progress is not to assume that we have arrived but to continually aim for the goal."
Bernard of Chiaravalle, Benedictine monk

Six years have passed since this book was printed and distributed for the first time. It was in English, but the Chinese version arrived just few years later.

Many changes have occurred in the world in this period, including some unexpected events as the experience of a pandemic spread. We thought that uncontrolled infections were historical events that occurred during the "dark ages" centuries ago. Once more, life and reality are more creative than humans.

In the smaller world of congenital interventional cardiology, several less dramatic changes have occurred. Our goal, as Bernard of Chiaravalle opined, remains the same and we are continually striving to achieve it. We all aim at improvement and at providing new answers to unsolved questions and problems.

As a result, we welcome and commend to you the second edition of this book. A good proportion of the content is new since new approaches and technologies continue to evolve. These include the transcatheter treatment of sinus venosus ASD, of PDA in premature babies, the use of self-expanding valves in large RVOTs, the development of engineering methods including modeling and 3D printing, the great development of imaging techniques, and the integration of imaging modalities to make procedures safer, quicker, more reproducible, and most importantly more effective. To these topics we have also added new chapters covering the approach to the mid-aortic syndrome, to PA-VSD-MAPCAs management, hybrid access, percutaneous solutions for large RVOTs with the current available technology, RVOT stenting in infants, and PFO closure. Furthermore, a chapter on tools and techniques usually used by "adult cardiology colleagues" was included in order to share knowledge about allied and potentially useful devices. Finally, as we are determined to strive for the best, a full and comprehensive chapter on quality evaluation and improvement has been included. Nowadays, excellence requires analysis of our own practice in order to continuously improve our results.

We are extremely proud that this book is the product of the experience of more than 50 world-class authors from all over the globe. It is thanks to them that this dream has come true … and for the second time!

A Roman philosopher, Seneca, said "Optimae ideae pertinent ad omnes" (the best ideas belong to everyone). We are grateful to each author for having shared their valuable experience and best ideas for the benefit of everyone and in particular for the benefit of our young colleagues and our patients.

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Part I

General

1 Patient Information and Informed Consent

Maarten Witsenburg

1.1 Introduction

Interventional (and diagnostic) catheterization is an important tool in congenital heart disease. It has evolved from atrial septostomy in the 1970s to a wide range of procedures including device closure of various defects and percutaneous valve implantation nowadays.

As any form of invasive study or treatment, it is not without risks and serious complications may occur. Therefore, it should only be performed after balancing the advantages and risks of the procedure [1]. The risk associated with the use of ionizing radiation for these procedures should also be kept in mind, especially because of the young age of many of these patients.

The patient (or his or her legal representative) has to agree on the suggested treatment, but can only do so after having been informed appropriately. The combination of the duty to inform and the agreement of the patient with the treatment plan is called informed consent.

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Informed consent is an essential step in any diagnostic or interventional cardiac catheterization in a patient with congenital cardiac disease.

1.2 Background

Healthcare ethics is based on the moral concepts of benevolence, autonomy, absence of malice, equity, and responsibility. Autonomy implies that the patient (and/or the legal representatives) can only consent after the provision of adequate information. The major elements in a valid consent process are sufficient understanding, sufficient information, and freeness from duress [2, 3].

In the ESC-EACTS myocardial re-vascularization guidelines, it is stated that information should be "objective and unbiased, patient oriented, evidence based, up-to-date, reliable, understandable, accessible, relevant, and consistent with legal requirements" [3].

1.3 Information and Consent in Clinical Practice

In a non-emergent setting, the indication for a diagnostic or interventional cardiac catheterization should be discussed within a multidisciplinary team including at least the (pediatric) cardiologist, interventional cardiologist, and cardiac surgeon. For noncomplex cases a written and locally approved protocol can be an alternative for the discussion within the multidisciplinary team.

In such a heart team, the indication, risks and benefits, possible other treatment options, and timing of procedure are discussed. This team decision is written down in the patient record, as well as the team members who were involved in the discussion.

Once the decision is made, the patient (or legal representative) is informed. It is important to take enough time to discuss the reason for treatment, its timing, risks, and possible treatment complications. Use of terminology that the patient understands is essential. An illustrated information sheet can be very helpful. One should realize that a lay person as a patient will always have a major lack of knowledge, even after an extensive discussion with the interventional cardiologist. The consent will therefore for a major part be based on the patient's trust in the treating physician. After the patient has consented, this is documented in the patient record. Depending on local rules and practice, the consent can be given orally or in writing.

In emergencies, time may be lacking to fulfill the steps mentioned above. A typical example is severely hypoxic neonate with d-transposition in need of urgent balloon atrial septostomy to improve atrial mixing. In such cases the information needs to be given after the procedure, including explanation of possible complications that may have occurred.

1.4 Conclusion

Recommendations for treatment in congenital heart disease will rarely have a higher than 1C level of evidence. As such expert opinion plays a major role. Even for procedures that have been used extensively for many years, the implications, including complications, have become clearer recently. The complex trajectory from indication for treatment to the diagnostic or interventional procedure itself are at risk for cognitive bias [4].

In addition the availability of an extending range of devices might sometimes result in using these for questionable indications.

The important point is that any interventional cardiologist should act in a responsible way before, during, and after the intervention. Whenever complications may have happened, he/she should be able to explain the problem, both to the patient and to colleagues, and how steps were taken to minimize any further harm.

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2 Anaesthesiological Management of the Paediatric Patient in the Catheterisation Laboratory

Giuseppe Isgrò and Marco Ranucci

2.1 Introduction

The widespread use of therapeutic cardiac catheterisation in the management of congenital heart disease requires the presence of a trained paediatric cardiac anaesthesiologist with the ability to provide both safe and consistent sedation or general anaesthesia to paediatric cardiac patients. Specific knowledge of the pathophysiology of congenital cardiac lesions and the clinical implications of diagnostic and therapeutic procedures are essential.

Sedation is often preferred to general anaesthesia, in particular for diagnostic procedures, because mechanical ventilation can cause haemodynamic disturbance and can alter the results of the study. General anaesthesia is applied mainly for interventional procedures (i.e. percutaneous valve implantation, atrial septal defect or ventricular septal defect closure, patent ductus arteriosus closure, or STENT implantation), during which it is essential to keep the patient deeply relaxed to permit the precise deployment of the device.

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Other factors influencing the decision to use sedation or general anaesthesia are patient related: age, clinical conditions and complexity of cardiac lesions.

Diagnostic and interventional procedures in the catheterisation laboratory carry risks for the patient such that continuous patient monitoring is essential.

The anaesthesiologist should contribute to the treatment of complications associated with cardiac catheterisation and, obviously, pre-empt and manage issues arising from sedation and anaesthesia. Finally good anaesthetic practice means that after the procedure the patient is delivered to a post-anaesthesia care unit or directly to the intensive care unit in the best condition possible.

2.2 Anaesthesia

2.2.1 Preoperative Consideration

Preoperative clinical evaluation is mandatory to assess the general condition of the patient and the type of cardiac disease and make plans for post-procedural care. Those patients affected by severe cyanosis should be hydrated prior to cardiac catheterisation, to minimise dehydration.

Fasting should be planned according to the age, clinical condition, and related laboratory investigations.

Routine preoperative tests (ECG, chest X-ray, lab investigations) are required and evaluated by the anaesthesiologist—in some cases, review of echocardiography. An assessment should include scrutiny of previous anaesthetic records and prior premedication.

Certain patients including chronically cyanotic patients are at risk of post-procedural bleeding, so that packed red cell units, fresh frozen plasma and concentrated platelet units are quickly available according to the procedure.

Strict attention to intercurrent illness is required, and if necessary, the catheterisation procedure should be postponed whilst this resolves.

2.2.2 Premedication

Drugs for premedication are administered to reduce anxiety and promote cooperation. Additional benefits include induction of anaesthesia without memory of this stressful time and reduced adrenergic stimulation that can be deleterious, particularly certain anomalies (i.e. tetralogy of Fallot, uncompensated ventricular septal defect with pulmonary hypertension and anomalous origin of left coronary artery arising from the pulmonary artery).

Children under 6 months of age or those that are very sick often can be managed without premedication as this can be deleterious under some circumstances.

Many drugs are available for premedication; the most commonly used are ketamine, midazolam, fentanyl and morphine. Dexmedetomidine, a new centrally acting alpha 2-adrenoceptor agonist, has been used in the setting of cardiac catheterisation laboratory safely with good results.

The choice of the drug alone or in combination must be decided by the anaesthesiologist after assessment of the patient and according to local experience and protocols.

2.2.3 Sedation and Anaesthesia

Sedation and general anaesthesia can be administered according to the preoperative condition, including the risk of developing post-procedural deleterious effects related to cardiac catheterisation and anaesthesia drugs use (i.e. pulmonary hypertension).

Pathophysiology of any cardiac lesion should be discussed beforehand with the paediatric cardiologist to reduce the risk of anaesthesia delivery, although modern anaesthesia drugs have reduced impact on cardiovascular system (Table 2.1). Sevoflurane, a volatile anaesthetic, has very little effects on systemic pressure and heart rate. Dexmedetomidine is thought to be protective for postoperative atrial fibrillation.

Table 2.1 Anaesthetic drugs

Drug	Induction	Maintenance
Ketamine	0.5–2 mg/kg	0.01–0.05 μg/kg/min
Midazolam	0.1–0.3 mg/kg	1–3 μg/kg/min
Propofol	1–2 mg/kg	3–5 mg/kg/h
Sevoflurane	3–5%	1–2%
Fentanyl	3–5 μg/kg	1–2 μg/kg/min
Morphine	0.1 mg/kg	1–2 μg/kg/min
Cisatracurium	0.1–0.2 mg/kg	1–2 μg/kg/min
Dexmedetomidine	1 μg/kg	0.2–1.4 μg/kg/h

Midazolam is safely used to maintain sedation, usually in combination with fentanyl or morphine.

The use of muscle relaxants that permit to keep the patient ventilated under general anaesthesia is nowadays safe, because the introduction of many newer agent with low rate of adverse effects; the combination of modern volatile anaesthetics and modern muscle relaxants have reduced to very rare event the incidence of malignant hyperthermia.

Cisatracurium, a non-depolarising muscle relaxant, a cis-isomer of atracurium, is indicated in paediatric anaesthesia because of the absence of histamine release; its half-life is 22–29 min and it is eliminated through the Hoffman metabolism, so it can be used safely in patients with poor renal function.

2.3 Monitoring and Anaesthetic Equipment in the Cardiac Catheterisation Laboratory

2.3.1 Electrocardiogram

Electrocardiogram is used for continuous monitoring of heart rate, rhythm and ST changes throughout the pre-, intra- and post-procedural phases.

2.3.2 Blood Pressure

Systemic blood pressure may be monitored noninvasively during the most common procedures by an automated oscillometric technique.

During risky procedures or in very sick patients, it may be necessary to monitor invasive blood pressure, by cannulation of an artery. Thereby arterial cannulas, transducers, and flushing devices must be present in the laboratory.

2.3.3 Pulse Oximetry

It provides a continuous and noninvasive monitor of oxygen saturation, which is mandatory during both sedation and general anaesthesia in paediatric cardiac patients, who are at risk for the development of hypoxia.

2.3.4 Capnometry

Is a continuous and noninvasive method of measurement of expired carbon dioxide and is very useful to monitor the adequacy of ventilation during general anaesthesia or to detect malfunction or failure of the anaesthesia machine. Moreover, it provides a useful information related to the quality of pulmonary perfusion and can reflect haemodynamic changes.

2.3.5 Temperature Monitoring

Temperature monitoring is very important especially in the newborns, who are at risk for hypothermia because of their relatively large surface area and the inefficiency of their thermoregulatory mechanisms. Cutaneous temperature may be monitored by adequate probes. Central temperature, if required, can be measured using a nasogastric probe.

In order to avoid hypothermia in children, it is important to warm the environment and the inhalatory gases by a humidifier. Warming of intravenous fluids may be needed. The use of heating blankets is also recommended especially in the newborn.

2.3.6 $ScvO_2$ (Continuous Mixed Venous Oxygen Saturation) Monitoring

Paediatric and adult patients with severe cardiac disease, who undergo catheter laboratory interventional procedures, can be monitored with respect to cardiac output. In congenital heart disease patients, it is usually either not possible or desirable to insert a pulmonary artery catheter designed for output measurement. Currently central venous catheters with oximetry are available to continuously monitor venous saturations. These catheters are usually inserted into the right jugular internal vein like a normal central venous line, with the same dimension and length (PediaSat and PreSep catheters—Edwards Lifesciences, Irvine, CO) (Fig. 2.1).

The continuous monitoring of venous saturation can help to identify sudden changes in haemodynamic status, rapidly changing when cardiac output decreases or increases.

This parameter is included also in the management of the early goal-directed therapy (EGDT) for critically ill patients.

2.3.7 NIRS (Near-Infrared Spectroscopy) Monitoring

Another tool of haemodynamic monitoring is near-infrared spectroscopy (NIRS) (Fig. 2.2). NIRS is used in many clinical situations to continuously monitor cerebral and splanchnic perfusion and has the potential to provide information on the adequacy of systemic oxygen delivery. Some authors have demonstrated a good correlation between NIRS and $ScvO_2$, but NIRS cannot precisely predict $ScvO_2$ value, though it can be used for trend monitoring.

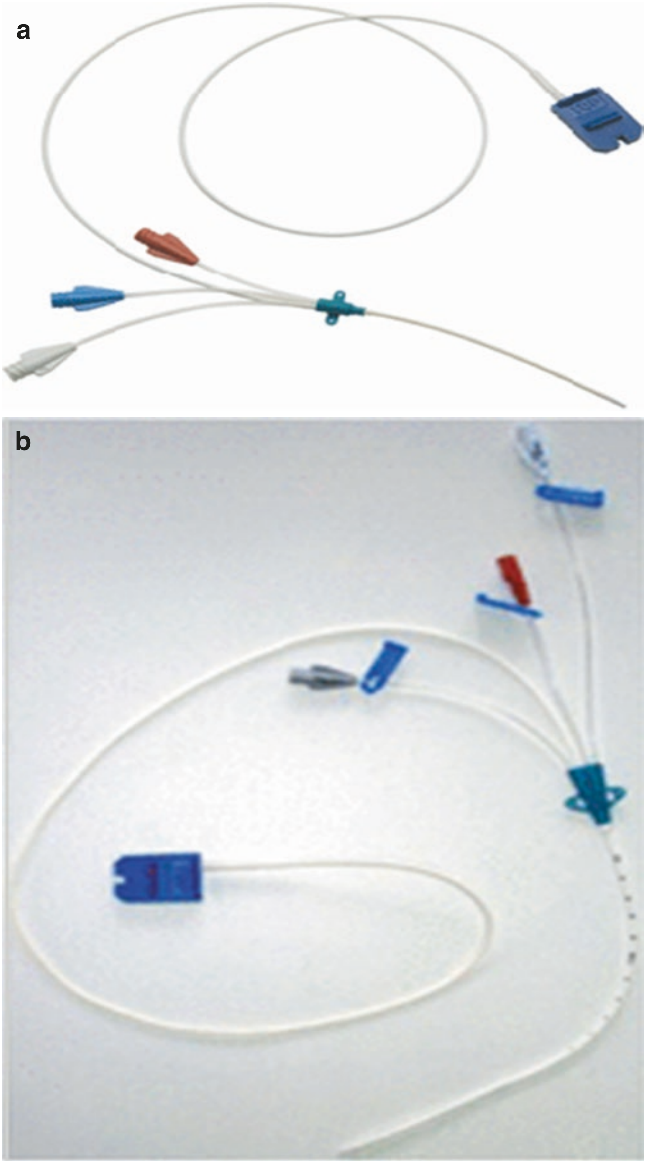

Fig. 2.1 (**a**, **b**) PediaSat catheter (paediatric) (**a**) and PreSep catheter (adult) (**b**). (Courtesy of Edwards Lifesciences)

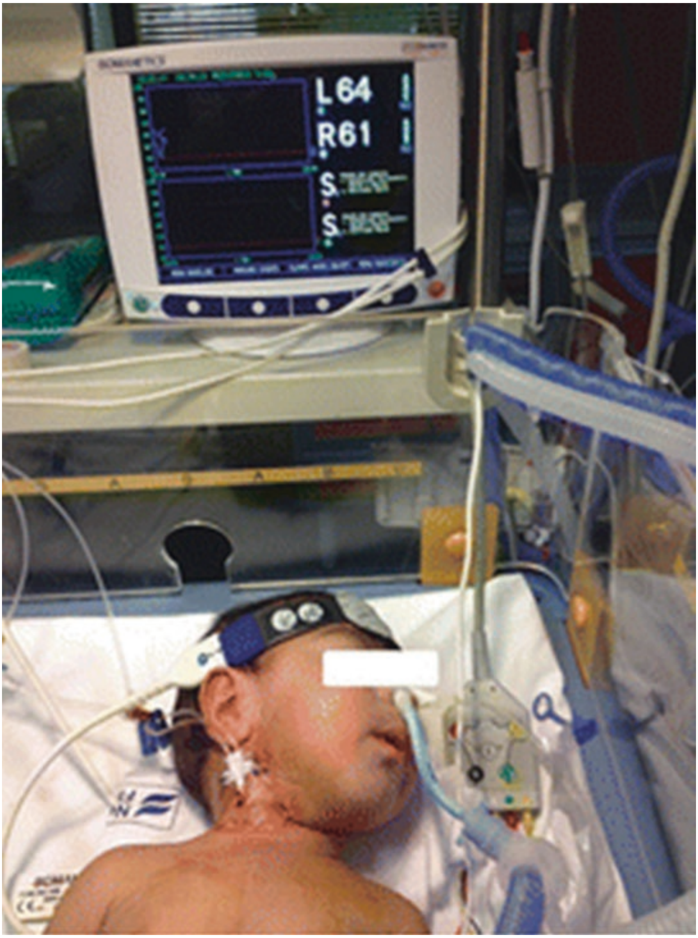

Fig. 2.2 NIRS monitoring

2.3.8 Anaesthetic Equipment

- Different sizes of cannulas for venous and arterial cannulation.
- Different sizes of central venous catheters.
- Different sizes of face masks.
- Different sizes of endotracheal tubes.
- Airway management equipment and difficult airway management equipment available.
- Suction apparatus and different sizes of suction catheters.
- Mechanical ventilator with inhalatory anaesthetic agents.
- Scavenging setup for waste inhalational agents.
- Sedative, analgesic and anaesthetic drugs.
- Resuscitation drugs.
- Intravenous infusion set—intravenous fluids (crystalloids and colloids).
- Defibrillator.
- Stethoscope.
- Self-inflating manual resuscitation bag.
- Foley catheters and nasogastric tubes.
- Blood gas analyser.

2.4 Ventilation Strategies During Cardiac Catheterisation Procedures

There is no preferred ventilation strategy in the cardiac catheterization laboratory, but spontaneous breathing, in particular during diagnostic procedures, is preferable to mechanical ventilation to maintain more natural intrathoracic physiology and consequentially to obtain the best acquisition of more accurate hemodynamic data.

General anaesthesia with positive pressure ventilation provides a secure airway and control of $PaCO_2$, but increased intrathoracic pressure may alter hemodynamic parameters.

However, general anaesthesia with positive pressure ventilation is not contraindicated, but requires a correct approach to the