Electrophysiology in Patients with Congenital Heart Disease

Sophia P. Poorsattar, Jeremy P. Moore, and Komal Patel

Abstract

Cardiac arrhythmias are among the most prominent source of morbidity, impaired quality of life, and mortality in patients with congenital heart disease (CHD). Rhythm disorders encountered in CHD patients span the entire spectrum of brady- and tachyarrhythmias while also ranging in symptomatology and clinical significance. Accordingly, many of these patients require diagnostic or therapeutic interventions in an electrophysiology laboratory. This chapter will discuss the mechanisms behind various arrhythmias, specific procedures performed for diagnosis and treatment, and procedural specific considerations specific to patients with CHD.

Keywords

Ablation · Arrhythmia · Conduction Defibrillator · Electrophysiology · Pacemaker

Introduction

Cardiac arrhythmias are among the most prominent source of morbidity, impaired quality of life, and mortality in patients with congenital heart disease (CHD) (Walsh and Cecchin 2007). This is especially true for patients who underwent surgical correction or palliation at a young age, only to present at a later date with an arrhythmogenic myocardium caused by their inherently atypical anatomy, surgical scars, and chronically remodeled heart after years of suboptimal hemodynamics. Rhythm disorders encountered in CHD patients span the entire spectrum of brady- and tachyarrhythmias while also ranging in symptomatology and clinical significance. While certain arrhythmias may be non-disruptive or benign, others may be poorly tolerated or life-threatening. As such, many of these patients require diagnostic or therapeutic interventions in an electrophysiology laboratory. An understanding of the mechanisms behind various arrhythmias, specific procedures performed for diagnosis and treatment, and procedural specific considerations will help to ensure safe and optimal anesthetic care for this unique patient population.

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The types of procedures that patients present for in the electrophysiology laboratory can be broadly divided into two basic categories: (1) electrophysiology study (EPS) and catheter ablation and (2) cardiac implantable electronic device (CIED) procedures.

Electrophysiology Study and Catheter Ablation in CHD

The spectrum of arrhythmias seen in the CHD population varies according to the underlying anatomical defect and the method of surgical repair or palliation. Arrhythmia pathophysiology is multifactorial and influenced by intrinsic cardiac anatomy, structural remodeling in the setting of abnormal pressure and volume loads, cellular injury and dysfunction related to low perfusion or hypoxic states, and tissue trauma and fibrosis at the sites of surgical interventions (Walsh 2007). With the wide-ranging types of anatomical defects and methods of surgical repairs or palliations, arrhythmias seen in the CHD population span the entire spectrum of tachyarrhythmias. While the preferred treatment options for certain rhythm disorders are pharmacologic or device therapy, for many tachyarrhythmias, EPS and catheter ablation are preferred. The most common indication for catheter ablation in the CHD population is recurrent supraventricular tachycardia, especially intra-atrial reentrant tachycardia (IART).

Atrial and Supraventricular Tachyarrhythmias

IART, which involves a macroreentrant circuit through abnormal atrial myocardium, is the most common arrhythmia mechanism in CHD patients. While IART can develop in the setting of nearly any congenital lesion, it is seen in up to 50% of patients with prior Fontan palliations and in approximately 30% of patients with prior Mustard or Senning operation (Walsh 2007). Fibrous tissue formation from surgical scarring, suture lines, or baffle insertion leads to the discontinuity of atrial muscle bundles and central obstacles for the development of reentrant circuits via anisotropic conduction. With atrial rates of 150–250 beats per min, 1:1 conduction can result in marked hemodynamic compromise or even complete circulatory collapse and are associated with a twofold increased risk for mortality (Khairy et al. 2014). As antiarrhythmic drug therapy is associated with poor long-term success in this condition, treatment has largely shifted to interventional procedures with acute ablation success achieved in 98% of patients (Moore et al. 2021).

As seen in those with typical anatomy, atrial fibrillation can develop in CHD patients that have conditions which result in marked dilation of the left atrium, such as in those with unrepaired atrial septal defects, left-sided valvular disease, and systemic ventricular dysfunction. While trials of cardioversion and pharmacologic rhythm control are often attempted first, catheter-based pulmonary vein isolation and rarely atrioventricular (AV) nodal ablations are alternative treatment options to surgical Maze procedures (Liang et al. 2019).

Accessory AV pathways, resulting in orthodromic AV reentrant tachycardia and AV nodal reentrant tachycardia, involve pathways that bypass or involve (in the case of AVNRT) the normal AV conduction pathway to connect the atrium and ventricle. In this case of antegradely conducting accessory pathways, this may lead to ventricular preexcitation, placing these patients at elevated risk for sudden cardiac death. Accessory pathways are found in a higher proportion of CHD patients with Ebstein's anomaly and levo-transposition of the great arteries (TGA) in which an Ebstein-like malformation exists suggesting a link between the AV valvular deformity and this conduction disturbance (Walsh 2007). In light of this well-appreciated association, catheter ablation has become the preferred treatment of this condition in those with symptomatic SVT or manifest preexcitation (i.e., Wolff Parkinson White pattern). For Ebstein's anomaly in particular, preoperative assessment of accessory pathways is often performed regardless of the symptoms or ECG findings, given the

high prevalence of occult arrhythmogenic substrate (Shivapour et al. 2014). However, mapping and ablative challenges exist in many patients due to displacement of the tricuspid valve hinge-point that occurs along with atrialization of the right ventricle in Ebstein's anomaly repair. As such, rates for ablation success and lack of arrhythmia recurrence for patients with Ebstein's anomaly are lower than in the general population (Khairy et al. 2014).

Guidelines described by the Pediatric and Congenital Electrophysiology Society (PACES) in conjunction with the Heart Rhythm Society (HRS) state that for atrial tachyarrhythmias in adults with CHD, catheter ablation is a Class I recommendation in those with recurrent symptomatic and/or drug-refractory IART or focal atrial tachycardia, those with recurrent symptomatic and/or drug-refractory supraventricular tachycardia related to accessory AV connections or twin AV nodes, and those with ventricular pre-excitation and high-risk or multiple accessory pathways. For patients with symptomatic drug-refractory atrial fibrillation, catheter-based ablation is a Class II recommendation (Khairy et al. 2014).

Ventricular Tachycardias

While ventricular ectopy and non-sustained ventricular tachycardia are common, sustained ventricular tachycardia is rare, occurring in only 0.1–0.2% of adults with CHD (Gallego et al. 2012). Of the CHD lesions, sustained monomorphic ventricular tachycardia occurs most commonly in tetralogy of Fallot where the right ventricular outflow tract is markedly scarred post-surgical intervention (Khairy et al. 2014). While sustained ventricular tachycardia is associated with well-described isthmuses for catheter mapping and ablation (Zeppenfeld et al. 2007; Moore et al. 2013), recurrence is possible even after acute ablation success. Currently, there is lack of consensus as to whether ICD implantation is required despite successful catheter ablation of monomorphic VT in the setting of tetralogy of Fallot and this is an area of active investigation. Such consideration should be made carefully and in the context of follow-up EPS to document non-inducibility after successful catheter ablation.

Society guidelines for catheter ablation of ventricular arrhythmias in adults with CHD provide a Class I recommendation for its use as an adjunctive therapy to ICD in those with recurrent monomorphic ventricular tachycardia, ventricular tachycardia storm, or multiple appropriate shocks that are not manageable by drug therapy or device reprogramming. Catheter ablation is a Class II recommendation in those with select ventricular tachycardias with high-risk features such as poor hemodynamic tolerance, deteriorating ventricular function, or as an alternative to adjunct drug therapy (Khairy et al. 2014).

Procedural Considerations

Transvenous EPS dates back to 1971 in which the first simultaneous electrical stimulations with recordings of intracardiac signals were performed (Wellens et al. 1972). Over time, advancements in EPS have allowed detecting tachyarrhythmia mechanisms and the localization of arrhythmogenic foci with improved precision with interventional catheter-based ablation techniques for the treatment of tachyarrhythmias emerging soon after. This improved understanding of rhythm abnormalities has proven especially useful when diagnosing and treating the complex rhythm disorders seen in the CHD population.

Electrophysiology studies and catheter ablation for patients with CHD must be performed in dedicated electrophysiology laboratories where procedural specific equipment is readily available. This includes external defibrillation capability, biplane fluoroscopy, an electrocardiographic and intracardiac electrogram recording system, an electroanatomic ("3D") mapping system, and readily available access to real-time laboratory assessment. Access to remote magnetic navigation (RMN) may serve as an additional resource in some situations. Due to their complex anatomy and physiology,

the American Heart Association/American College of Cardiology recommend that these procedures be performed by electrophysiologists with expertise in the management of adult CHD (Stout et al. 2019). Similarly, it is advisable that these procedures be performed at specialized centers with anesthesiologists and cardiac surgeons with an expertise in the care of adult CHD patients (Finnerty and Griffin 2021). In patients with CHD necessitating structural intervention, there has also been an emergence of specialized centers performing concomitant electrophysiologic and structural interventions with favorable outcomes (Lindsay et al. 2018).

While the femoral vein is the most common choice for venous access, alternative approaches may be required based on patient anatomy, including subclavian, internal jugular, or transhepatic (e.g., with interrupted IVC or femoral venous occlusions). If the procedure requires entry into the left side of the heart, the approach is generally transseptal, transbaffle/transconduit (Moore et al. 2020), or even transpulmonary (Moore et al. 2016a). Less commonly, a retrograde aortic approach is utilized (especially when using RMN).

Once access is obtained, various mapping techniques can be utilized to identify sites for catheter ablation. For organized and hemodynamically tolerated tachyarrhythmias, three-dimensional electroanatomic mapping is generally performed first to delineate the key components of (or even the entire) reentrant circuit. The map is then evaluated to determine the optimal catheter ablation site that is critical to tachycardia maintenance (Fig. 1). When choosing the optimal ablation target, care is required to avoid potential collateral damage to structures, such as the phrenic nerve, sinus or AV node, or coronary arteries. Prior to catheter ablation energy delivery, entrainment is typically performed at the potential target site to verify critical participation in the reentrant circuit.

The use of three-dimensional (3D) mapping systems has allowed for improved mapping and ablation success in the complex targets observed in CHD. Electroanatomic mapping systems allow for a 3D reconstruction of electrograms and chamber geometry. Visually, this takes the form of color-coded static maps depicting isochrones of electrical activity merged with cardiac reconstructions from magnetic resonance or computed tomography imaging obtained prior to the procedure (Walsh 2007; Chua et al. 2012). Recognizing the improved success with this mapping technique, society guidelines recommend that EPS for adults with CHD be performed at facilities with this technology available (Khairy et al. 2014; Stout et al. 2019).

Upon identification of the critical isthmus of the reentrant circuit, catheter ablation is performed to render it electrically inert via either irrigated radiofrequency (burning) or cryothermal (freezing) energy (Chua et al. 2012). With radio frequency catheter ablation (RFA), the catheter tips produce high tissue temperature through resistive (and to a lesser extent, conductive) heating to disrupt the tissue and create electrical scar. In cases where proximity to the AV conduction system is expected ("peri-nodal substrates"), cryoablation may be the preferred energy source over radiofrequency energy in CHD (Khairy et al. 2014).

Anesthetic preparation for EPS and catheter ablation starts with a pre-anesthesia assessment, which should include the arrhythmia to be addressed, the patient's anatomy and cardiovascular reserve, and the anticipated procedural approach including if a concomitant procedure is to be performed. The choice of anesthetic technique is partly dictated by patient comorbidities, length of procedure, and anticipated complications. In those with limited cardiopulmonary reserve, vasoactive support, advanced hemodynamic monitoring, controlled ventilation, and frequent lab checks may be required. With technically challenging cases, procedural times can exceed 4 h in duration during which the patient is expected to lay flat and remain motionless to facilitate accurate mapping and ablation. Placement of foley catheter should be considered for these prolonged complex procedures.

The type of ablation procedure also influences anesthetic management. In patients undergoing supraventricular or ventricular tachycardia ablation, a light sedation technique may be preferred

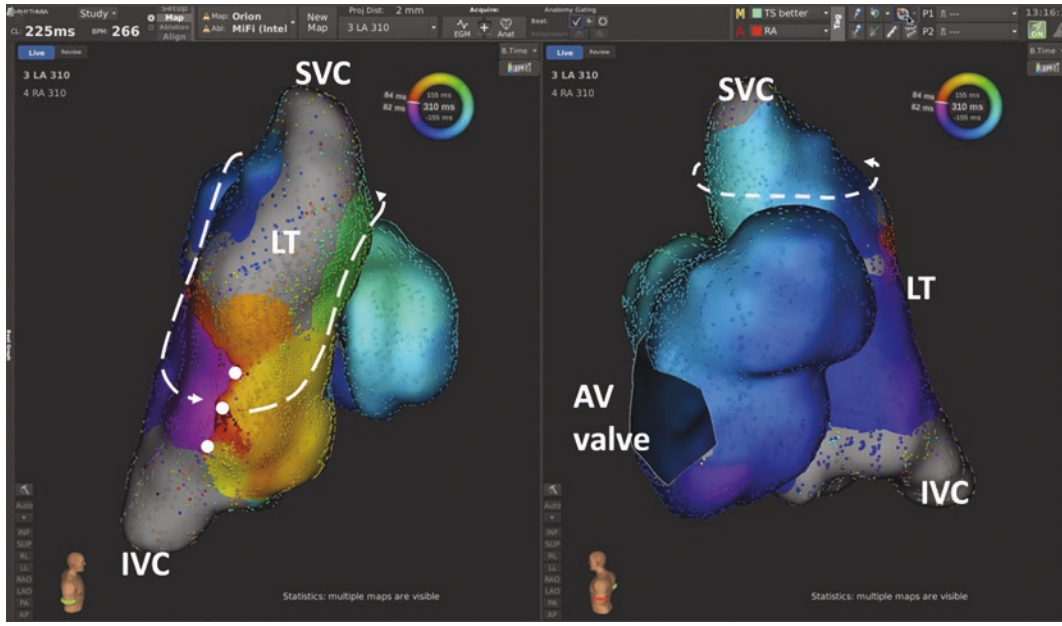

Fig. 1 Example of a three-dimensional electroanatomical map in a patient with recurrent intra-atrial reentrant tachycardia after lateral tunnel Fontan operation (right and left lateral views are shown). There is a macroreentrant circuit (atrial rate 190 beats per min; dashed white arrows) that rotates around a combined obstacle incorporating the SVC and morphological right atrial atriotomy. The circuit was successfully interrupted with irrigated radiofrequency energy applied to the isthmus between the lower end of the atriotomy incision and the IVC (solid white circles). *AV* atrioventricular, *LT* lateral tunnel, *IVC* inferior vena cava, *SVC* superior vena cava

to avoid suppression of the ventricular arrhythmia. In patients undergoing atrial fibrillation or atrial flutter ablation, a transesophageal echocardiogram may be required to rule out intracardiac thrombus prior to proceeding with the case. In atrial fibrillation ablations, a general anesthetic technique is performed with placement of an orogastric tube and esophageal temperature probe for reducing the rare but possibly fatal complication of atrio-esophageal fistula formation. Additionally, neuromuscular blockade is avoided to allow for recognition of phrenic nerve stimulation from the ablation catheter when targeting either the right sided pulmonary veins or posterolateral right atrium. With transseptal puncture for any ablation procedure, systemic heparinization is required to reduce the risk of thromboembolism. Caution must be taken with fluid management as the electrophysiologist may use 2–3 L of crystalloid while irrigating the ablation catheters or a transseptal sheath throughout the case. During mapping, the patient may experience frequent rhythm changes, which may result in hemodynamic instability and necessitate cardioversion or defibrillation. In the event of inadvertent cardiac perforation and pericardial tamponade, the electrophysiologist may have to place a pericardial drain. With local anesthetic infiltration at the cannulation sites, postoperative pain is usually minimal and is effectively managed with intravenous or oral analgesics as needed. Disposition to the postanesthetic care unit or intensive care unit depends on the patient's hemodynamic condition and the complexity of the procedure.

Cardiac Implantable Electronic Devices

CIEDs fall into two principal categories, implantable cardioverter-defibrillators (ICDs), which are for the prevention of sudden cardiac death (SCD), and pacemakers, which are for the treatment of symptomatic bradycardia.

Implantable Cardioverter-Defibrillators

Sudden cardiac death is responsible for 20–25% of all-cause mortality in adults with CHD with the majority of causes being of arrhythmogenic etiology (73–80%) (Khairy et al. 2014). The congenital lesions identified to be highest risk for the development of SCD include tetralogy of Fallot, TGA (both dextro- and levo-variations), Ebstein's anomaly, left-sided obstructive lesions, and Eisenmenger syndrome (Silka et al. 1998; Koyak et al. 2012).

ICDs are placed for the primary or secondary prevention of SCD. As described in the 2014 PACES/HRS guidelines, Class I indications for the primary prevention of adults with CHD include those who meet standard recognized criteria (biventricular physiology with a left ventricular ejection fraction of ≤35% and advanced heart failure symptoms) as well as those with spontaneous sustained ventricular tachycardia. ICD therapy is also recommended to be considered in those with tetralogy of Fallot with high-risk features for SCD, cardiomyopathies or univentricular hearts with high-risk features for SCD, and syncope of unknown origin with known or suspected history of ventricular arrhythmia. As expected, ICD therapy for secondary prevention in those with a history of life-threatening arrhythmia is a Class I indication for adults with CHD (Khairy et al. 2014).

Traditional transvenous ICD (TV-ICD) placement mirrors that of transvenous permanent pacemakers (TV-PPM) with the subcutaneous or submuscular insertion of a pulse generator, normally in the left infraclavicular area, and leads that are affixed to the endocardium via a transvenous route, typically the subclavian vein. Among all patients, early complications of TV-ICD placement include vascular injury, pneumothorax, hemothorax, cardiac perforation, pericardial effusion, and tamponade. Late complications include lead failure and lead-related infective endocarditis (Bowman et al. 2021). Compared to the general population, TV-ICDs present many challenges in the adult CHD population as these patients often have complex central venous anatomy that is not amenable or potentially hazardous to device placement, such as venous anomalies, intracardiac shunts, Fontan anatomy, or mechanical right AV valves (Khairy et al. 2006). As an alternative to transvenous lead implantation, subcutaneous ICDs or an epicardial approach may be considered in these situations (also discussed below with pacemakers).

Due to the increased or prohibitive risk of TV-ICD placement in certain adults with CHD, novel configurations of ICD therapy have been developed. One such configuration is the subcutaneous ICD (S-ICD), which gained Federal Drug Administration (FDA) approval in 2012. Like a TV-ICD, a S-ICD consists of a generator and a shocking lead. With the S-ICD, however, the generator is inserted subcutaneously in the anterolateral position while the shocking lead is tunneled midline and with the coil oriented vertically in the left or right parasternal position (Fig. 2a, b). For adult CHD patients, S-ICDs offer many advantages. First, the superficial position outside of the thoracic cavity means vascular access is not required. This provides an attractive ICD therapy option in patients with no venous access, acquired stenosis or obstruction of central veins, awaiting heart transplantation, or prior and/or at risk for endovascular lead infection (De Maria et al. 2015). Further, in patients who have device failure or young patients who necessitate multiple device revisions throughout their lifetime, complications related to repeated transvenous lead additions or extractions are avoided (Brunner et al. 2014). Accordingly, S-ICD use is recommended for adult CHD patients when feasible (Al-Khatib et al. 2018; De Maria et al. 2014). Certainly, without endomyocardial lead insertion, limitations exist. S-ICDs have no pacing capabilities (aside from short-term post-shock back up pacing), a larger generator, a shorter battery life, and a prolonged time to therapy as compared to TV-ICDs. Additionally, certain patients may not be a suitable candidate for S-ICDs due to inadequate sensing of subcutaneous signals (such as patients with heavy weight, hypertrophic cardiomyopathy, prolonged QRS interval, or inappropriate R to T wave ratio) or anatomic characteristics that increase complication risk (such as insufficient subcutaneous tissue or chest wall abnormalities) (De Maria et al.

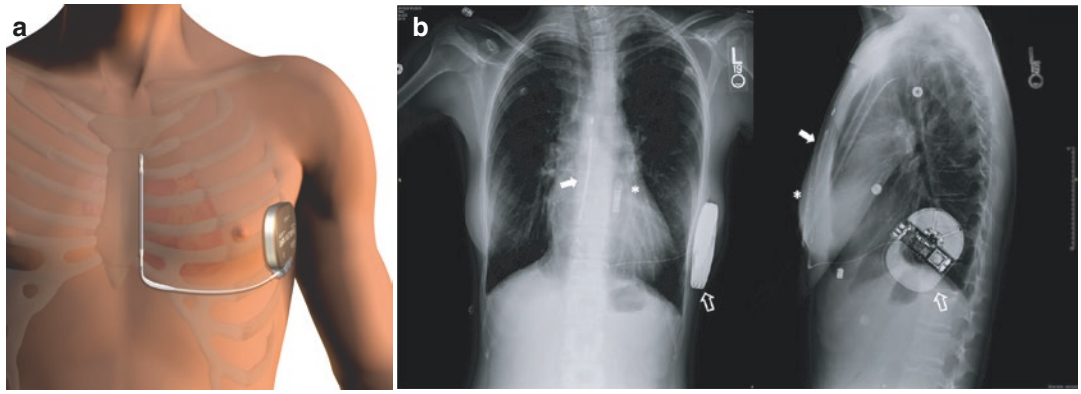

Fig. 2 (**a**) Schematic presentation of the of the anatomical placement of the Emblem™ Subcutaneous Implantable Defibrillator (S-ICD). (© 2020 Boston Scientific Corporation or its affiliates. All rights reserved. Used with permission from Boston Scientific Corporation). (**b**) Posterior-anterior and lateral chest x-ray after S-ICD placement in a woman with Eisenmenger syndrome and documented VT. The pulse generator has been placed in a left axillary position across from the apex of the heart (open arrowhead) with the subcutaneous shocking coil tunneled over the sternum (solid arrowhead). A prior implantable loop recorder is shown in a parasternal position (asterisk). Given the patient's congenital physiology, the patient underwent preoperative truncal plane block to limit the need for general anesthetic

2015). The safety and efficacy of the SICD in the CHD population has been previously described (Moore et al. 2016b).

Anesthetic considerations for TV-ICD placement mirror those of the considerations for TV-PPM implantation (also discussed below). For S-ICD placement, the decision for general versus sedation technique and types of monitoring depends on individual patient factors. The patient is positioned supine with the left arm abducted in order to facilitate exposure of the left axilla and chest wall. If performed under sedation rather than general anesthesia, adequate analgesia may be challenging with local lidocaine infiltration alone as the sizable generator box is inserted in an intramuscular position between the serratus anterior and the latissimus dorsi muscle and the lead tunneled through substantial subcutaneous tissue to reach its final parasternal position. To obviate the need of supplemental analgesics, regional blockade with a serratus anterior plane block may be effective for analgesia in the T2–T9 dermatomal distribution through blockage of the lateral cutaneous intercostal nerves (Droghetti et al. 2018). Once the device is inserted, one or more defibrillation threshold tests will be performed. This energy delivery may be quite unpleasant for an awake or minimally sedated patient, and a short-acting anesthetic agent is recommended at this point for patient comfort. As an energy delivery may lead to hemodynamic instability, in high-risk patients, this test may be avoided (Finnerty and Griffin 2021).

Pacemakers

Permanent pacing is often required in patients with CHD for bradyarrhythmias either due to a diseased conduction system (whether inherent to the anatomic substrate at birth or with progressive deterioration over time) or conduction system injury secondary to surgical or procedural interventions. Progressive, spontaneous AV block, for instance, is classically seen with levo-TGA, where the incidence approaches 2% per year postnatally (Graham et al. 2000; Beauchesne et al. 2002). The PACES/HRS guidelines describe the Class I recommendations for permanent pacing in adults with CHD to include those with symptomatic bradycardia from sinus node dysfunction or any degree of AV block, congenital complete AV block with ventricular conduction or function abnormalities, and post-operative high-grade second- or third-degree AV block that is not expected to resolve. Additional Class II indications include adults with CHD with vary-

ing degrees of sinus node or AV conduction disturbance with concerning features and for those with a high proclivity for developing IART (Khairy et al. 2014).

The two most common configurations for permanent pacemaker placement are transvenous or epicardial systems. Transvenous systems account for the majority of implants, while epicardial systems are reserved for patients not amenable to transvenous implantation. As mentioned previously, with TV-PPM placement, a pulse generator is typically placed in the left upper pectoral region with leads advanced to the endocardium via a transvenous route. With epicardial pacemaker implantation, surgical exposure of the heart is required. The choice of optimal generator position and route depends on various patient-specific factors.

The challenges of TV-PPM placement in the adult CHD population mirror those of TV-ICDs. Additionally, TV-PPMs are fraught with similar early and late complications related to the leads, generator pocket, and patient anatomy. To circumvent these issues, leadless pacemakers were developed, which combine a pacing lead and generator into one small device that inserts into the right ventricular septum via a transvenous approach (Fig. 3a, b). While two leadless pacemakers exist, Nanostim™ (St Jude Medical, Sylmar, CA, USA) and Micra™ (Medtronic, Minneapolis, MN, USA), presently, only the Micra™ is commercially available. In its current form, the device is only suitable in patients who require single right ventricular chamber pacing. While this includes a smaller pool of suitable candidates than TV-PPMs, leadless pacemakers are an attractive alternative in adult CHD patients with poor venous access in the upper body in whom permanent transvenous leads would be unfavorable. Further, at end of life, these devices offer the option of either endovascular device retrieval (if recently implanted) or abandonment with implantation of additional devices simultaneously without significant hemodynamic compromise (Finnerty and Griffin 2021). As the technology is relatively new, long-term durability and outcome data remain unknown; however, early complication rates are comparable to those seen with traditional TV-PPM (4.8% and 4.1%, respectively) (Tjong and Reddy 2017).

Anesthetic preparation for pacemaker placement starts with a pre-anesthesia assessment,

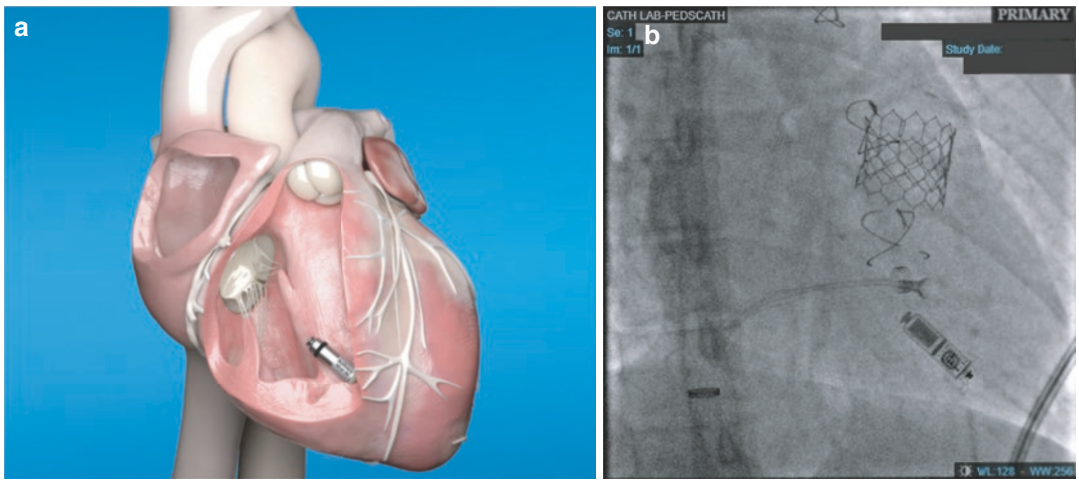

Fig. 3 (**a**) Schematic presentation of the anatomical placement of the Micra™ Transcatheter Pacing System. (Image courtesy of Medtronic). (**b**) Fluoroscopy during deployment of a leadless transcatheter pacemaker in a young woman with tetralogy of Fallot and intermittent episodes of bradycardia. The delivery system is shown in the body of the right ventricle with the pacemaker recently deployed at the septal right ventricular apex. The delivery sheath can be seen at the junction of the right atrium and the inferior vena cava. A transcatheter valve had been placed in the pulmonary position at the same procedure prior to transcatheter pacemaker placement in order to treat pulmonary stenosis

which should include an accurate understanding of the indication for pacemaker placement, the patient's anatomy and cardiovascular reserve, and the anticipated procedural approach. For TV-PPM, placement of a left upper extremity peripheral intravenous line allows for a venogram to be performed prior to incision. While important for all patients undergoing TV-PPM placement, this is especially pertinent in CHD patients with numerous congenital and post-surgical structural variances to confirm venous anatomy and patency. The choice of anesthetic technique and advanced monitoring are dictated by patient comorbidities. In the absence of contraindications, sedation with local topicalization in the area of the generator pocket and venous exposure is often well tolerated as this is most stimulating part of the procedure (Chua et al. 2012). With implantation of right atrial and coronary sinus leads, phrenic nerve capture is assessed and paralytics should be avoided. Using a sedation technique with oxygen administration via nasal cannula or face mask, precautions must be taken to reduce the risk of airway fire as the generator pocket is often implanted in the upper chest in close proximity to the airway with the use of electrocautery. To prevent oxygen accumulation under the sterile drape, an oxygen-air blender that can deliver a low concentration of oxygen and a supportive bar that can lift and secure the drape away from the face should be utilized. For leadless pacemaker implantation, venous access is normally percutaneously via the femoral vein. In contrast to TV-PPM placement, no chest wall dissection is required making the procedure much less stimulating and tolerable for most patients with minimal sedation and local anesthesia alone.

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