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REVIEW ARTICLE

Perioperative implications and management of dextrocardia

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Abstract Dextrocardia, a term used to describe all varieties of developmental malformations resulting in the positioning of the heart in the right hemithorax, is linked to a number of highly significant cardiac disorders. Current estimates vary tremendously in the literature. Only about 10 % of patients with diagnosed dextroversion show no substantial cardiac pathology; however, the incidence of congenital heart defects associated with dextrocardia is close to 100 %. The majority of studies previously reported include dextrocardia associated with situs inversus and cases of Kartagener syndrome. There is complex embryology and pathogenesis that results in dextrocardia. Physical examinations of the heart, such as percussion and palpation during routine exams, are vitally important initial diagnostic instruments. X-ray, CT scan, echocardiography (ECHO), and MRI are all invaluable imaging modalities to confirm and classify the diagnosis of dextrocardia. In summary, heart malposition is a group of complex pathologic associations within the human body, rather than just a single congenital defect. Clinicians such as anesthesiologists have unique challenges managing patients with dextrocardia. An appreciation of associated pathogenesis, appropriate diagnosis, and management is paramount in ensuring the best outcome for these patients perioperatively.

Keywords Kartagener syndrome · Dextrocardia · Heart looping · Situs inversus · Anesthetic management

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Introduction

Dextrocardia is an umbrella term which embraces all varieties of developmental malformations resulting in the positioning of the heart in the right hemithorax, with the base and apex of the heart pointing caudally and to the right. Dextrocardia refers strictly to abnormalities of embryologic origin that are intrinsic to the heart itself, rather than to those caused by extracardiac problems. The latter are referred to as cardiac dextroposition, which is usually a result of underlying lung pathology and other mediastinal structural abnormalities, causing the heart to deviate to the right. Pneumoectomy, right lung hypoplasia, diaphragmatic hernia, and pneumomediastinum are a few of the conditions that may result in heart dextroposition [1].

The major clinically relevant concern from dextrocardia is its link to congenital and acquired heart defects, as well as other organ-related abnormalities. This link depends strongly on the specific subtype of dextrocardia. However, disagreement between authors who have attempted to classify different types of dextrocardia, as well as the relative rarity of the condition, has resulted in limitations in our overall knowledge base. Unfortunately, the available research describing heart malposition confounds rather than assists clarifications. Moreover, data are lacking in the anesthesiology literature, other than a limited number of case reports. Furthermore, these reports primarily describe patients with Kartagener syndrome, a disease associated

with dextrocardia predisposing to airway pathology, but do not discuss other significant dextrocardia syndromes [2–5].

In this article, the existing literature on dextrocardia is reviewed. We also describe anesthetic approaches to a patient with suspected heart malposition. Another goal of this review is to demonstrate the most important features of airway and cardiovascular system anatomy and morphology, including stereoscopic cardiac chamber position, anatomy of the great vessels and their immediate branches (including coronary arteries), as well as other organs and systems that have implications for anesthesia. Finally, we describe the appearance of common cardiac and extracardiac disorders associated with particular types of dextrocardia which anesthesiologists may be asked to manage in clinical practice. Our review attempts to summarize the newest clinical findings and best synthesized practice strategies, focusing on diagnosis and perioperative management of patients with dextrocardia, in an extensive review for the clinical anesthesiologist.

Epidemiology

Since dextrocardia was thought not to be widely prevalent or clinically significant, it never drew significant attention from clinical practitioners. However, this is no longer the case. With increasing numbers of computed tomography (CT) scans and surgical procedures, there has also been an increasing number of incidental findings of dextrocardia. Additionally, after the introduction of new complex heart lesion repair techniques in the 1970s, approximately 85 % of patients who suffer from congenital heart diseases are now expected to survive into adulthood. There is ample reason for the claim that more and more people with residual cardiac lesions will be seen in operating rooms and intensive care units in the future. It has been reported in recent years that there are currently approximately 800,000 adults with treated congenital heart abnormalities [6, 7]. This number includes a proportion of patients with dextrocardia. Thus, we may currently expect to see not just asymptomatic patients diagnosed with cardiac malposition but also those with corrected dextrocardia-associated defects, which may have unknown long-term sequelae.

However, with the increased survival of children with congenital heart disease (CHD) in both normocardia and dextrocardia groups, it is a challenging task to obtain the true and precise incidence of dextrocardia and related malformations. This could be partially explained by the vague clinical significance of isolated dextrocardia and a huge proportion of patients with undiagnosed dextrocardia in an asymptomatic population. Nevertheless, there have been several attempts to estimate the occurrence of dextrocardia in the general public. Reported statistics therefore may not reflect the actual state of affairs, since the range of reported results is very broad (most likely due to lack of statistical power and the limited number of subjects involved). Estimates have shown that right thoracic heart can be found in 1 in 8000 people. One of the few retrospective studies that analyzed all congenital dextrocardia cases at a tertiary care hospital from 1985 to 2001 showed the incidence of dextrocardia to be 1 in 12,019 pregnancies [8]. This data was reported by Walmsley et al. [9] who evaluated 5539 fetal echograms and diagnosed 85 cases of dextrocardia, thus claiming the incidence of dextrocardia to be as high as 0.84 %. It is worth mentioning that this study was carried out in tertiary care centers for high-risk perinatology and all screening procedures were performed for standard indications [10], including any suspected heart pathology. Another analysis of fetal dextrocardia appears to suggest an overall incidence of 0.22 % [11]. In this study, a total of 36,765 pregnant women were screened for possible fetal heart problems, and again echocardiography (ECHO) was utilized as a screening method. While the incidence of fetal heart malpositions is relatively easy to assess, most studies report the incidence of dextrocardia in high-risk groups with anticipated heart problems. Indeed, obtaining the frequency of the pathology in adults might be an even more challenging task, due to the broad spectrum of manifestations that dextrocardia can have, as well as the high number of asymptomatic/undiagnosed patients. One of the few studies that were conducted show the disorder occurrence as 1 in 309; however, this was among all cardiac conditions. The authors of the aforementioned study reported 139 cases of dextrocardia out of a total of 42,950 patients who attended a cardiology clinic for any cardiac condition [12]. Although it might be argued that this condition is not as frequent in the general population, among those with some type of cardiac condition dextrocardia seems to be more common than originally thought.

In attempt to break down the range of dextrocardia disorders into different subtypes in order to categorize the clinical importance of individual variants, we found that the majority of studies reported in the literature included dextrocardia associated with situs inversus (and particular cases of Kartagener syndrome). This type of dextrocardia tends to be most prevalent and clinically significant among the wide range of dextrocardia disorders. The incidence of situs inversus was estimated to be 1 in 16,000 births [13]. Along similar lines, Brueckner et al. [14] argues that situs inversus may be found in up to 1 in 8500 patients in the general population. As for other dextrocardia related syndromes, the data is less evident and more inconsistent. Most existing case reports reveal only particular aspects of other dextrocardia related syndromes and include little epidemiologic data.

Pathogenesis and associated malformations

Heart malposition is a group of complex pathologic associations within the human body, rather than just a single congenital defect. The actual presentation of dextrocardia should alert medical practitioners to possible associated defects, some of which can be challenging. However, there are a couple of other substantial potential anomalies that may draw the attention of a clinician and, in particular, an anesthesiologist. These include airway disorders attributed to Kartagener syndrome, or anatomic disorders related to blood vessels and/or nerves.

Situs

Situs is a key component in understanding the disorders associated with dextrocardia, which may help to identify underlying assorted clinical manifestations. The concept of situs concerns the arrangement of structures within the human body. Cardiac malpositions are generally divided into 3 subtypes, depending on the presence of other organ asymmetry. Situs solitus describes the normal anatomy of organs found in the majority of the population, with the heart on the left, liver on the right, etc. Two alternatives of the normal viscera anatomy are situs inversus and situs ambiguous. Figure 1 illustrates the differences between situs solitus, situs inversus, and situs ambiguous.

As previously mentioned, there are several possible variations of dextrocardia, based on the situs presentation and the particular stage of development at which the malformation has occurred. Knowing the layout and intrinsic anatomy of the organs, heart chambers, and great vessels, one can anticipate underlying manifestations of heart malposition. Figure 2 depicts cardiac chambers and structures in dextrocardia. The mainstay of correct diagnosis is identification of the alignment of the heart and other organs. However, the systemic approach, which is discussed in greater detail in the diagnosis section, should be utilized to recognize interposition of heart structures, which will lead to more precise estimation of possible defects and risks [15]. We have made an attempt to summarize and systematize most common abnormalities seen in patients with heart malposition and the respective anatomic linkages. Patterns of cardiac and non-cardiac lesions will be discussed according to the situs type.

Situs solitus

Dextrocardia with situs solitus (dextrovesion) describes normal configuration of the viscera, with the liver on the right, spleen on the left, and normal positioning of the lungs. The only misplaced structure is the heart, which is located in the right hemithorax. The base-to-apex axis of the heart points to the right. It is the second most common configuration among the dextrocardia subtypes. If the intracardiac connections are concordant, the morphologic orientation of the heart chambers corresponds to the anatomic orientation; however, the right heart chambers are posterior to the left heart chambers. Dextroversion has a high rate of association with discordant atrioventricular (AV) connections and other cardiac anomalies—the other viscera tend to remain unaffected [12, 16, 17]. Only about 10 % of patients with diagnosed dextroversion show no substantial cardiac pathology [18]. However, previous studies showed the incidence of CHD associated with dextrocardia to be even higher, close to 100 % [19, 20]. Common cardiac malformations typically linked to dextroversion are: anomalous pulmonary venous return, tetralogy of Fallot, septal defects, coarctation of the aorta, corrected and uncorrected transposition of the great arteries (TGA), persistent ductus arteriousus, ventricular septal defect, pulmonic stenosis, and others. Although there is a high incidence of all previously listed CHD, the incidence of tetralogy of Fallot remains debatable. One study described 7 cases of tetralogy of Fallot linked to dextroversion [21]. However, other studies reported virtually no cases of tetralogy of Fallot associated with situs solitus dextrocardia, thus proposing it to be no more prevalent than in the general population [20, 22, 23].

Newborn cases of dextroversion usually suggest a huge variety of underlying pathologies, and may present with a wide range of defects. Severe abnormalities in newborns may require prompt surgical intervention, whereas adults with incidental findings of dextrocardia usually demonstrate little room for corrective measures. However, thorough cardiologic evaluation is still recommended for every patient newly diagnosed with dextroversion [24]. The major concern in this patient group in terms of frequency of anticipated disorders is the association with disrupted core heart anatomy. Any case of misplaced heart must be carefully assessed for atrial, ventricular, and arterial concordance. Double-outlet ventricles, double-inlet ventricles, a single ventricle, or AV discordance with corrected and uncorrected TGA are most common. Due to the relatively high rate of discordant atria and ventricles, dextroversion is associated with congenitally corrected TGA in 30–40 % of all cases [12, 25, 26]. While ccTGA may be attributed to normal hearts, nearly 20 % of all patients diagnosed with ccTGA have underlying dextracardia with situs solitus [27]. As discussed above, the problem seen with the correction is that the morphologic right ventricle and tricuspid valve cannot tolerate systemic pressures over time. Clinically significant regurgitation occurs in 20–50 % of patients. It may resemble mitral regurgitation in otherwise healthy patients, but progresses more rapidly

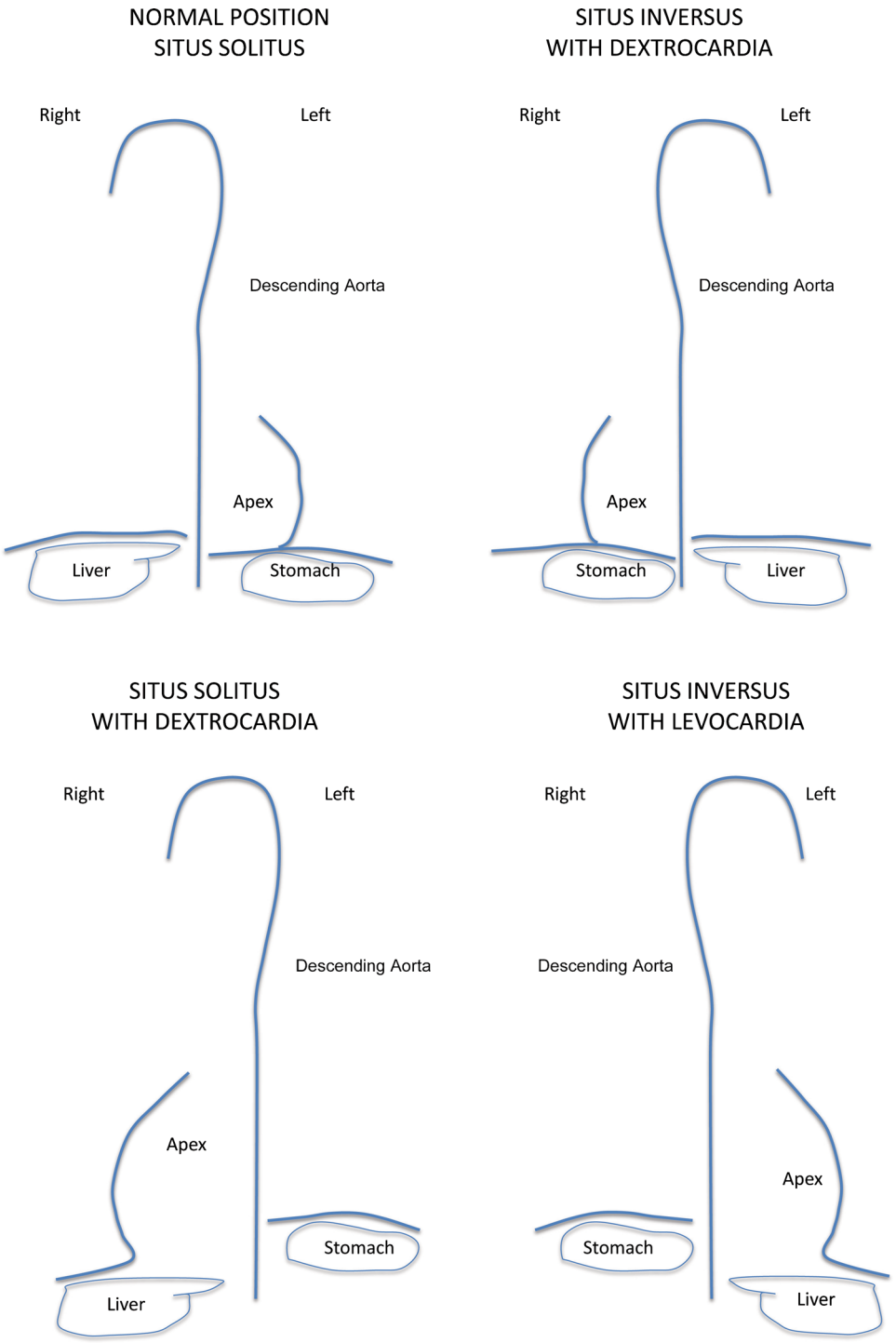

Fig. 1 Schematic illustrations of the four basic cardiac positions (normal and three malpositions) and the relationships of the descending aorta, cardiac apex, stomach, and liver as viewed on a frontal plain chest X-ray. In situs solitus, the descending aorta, cardiac apex, and stomach are on the left. In situs inversus with a right thoracic heart, the descending aorta, cardiac apex, and stomach are on the right. In situs solitus with a right thoracic heart, the descending aorta and stomach are on the left, and the cardiac apex is on the right. In situs inversus with a left thoracic heart, the descending aorta and stomach are on the right, but the cardiac apex is on the left. Reproduced with permission from Perloff [74]

[28]. While dextroversion may present with cardiac defects regardless of the concordance of heart chambers, existing ccTGA shows increased predisposition to particular cardiac anomalies. Systemic AV valve dysfunction, ventricular septal defect, and pulmonary outflow obstruction are commonly seen with congenitally corrected TGA [29]. Ventricular septal defect (VSD) is reported to be associated with ccTGA in 60–80 % of cases, pulmonary stenosis (PS) in 30–50 % [27]. Though cardiac discordance with associated ccTGA is a profound heart defect affecting the blood circulation, in rare cases when there is no additional heart lesion ccTGA may go undiagnosed until adolescence or even adulthood. While remaining relatively asymptomatic under normal circumstances, an event like surgery may act as

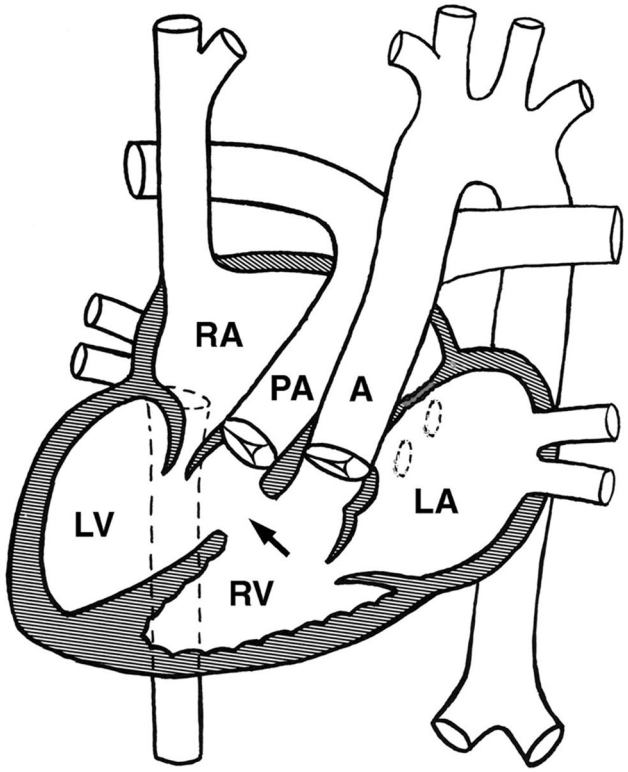

Fig. 2 The left atrium (*LA*) connects to the morphologic right ventricle (*RV*), and the right atrium (*RA*) connects to the morphologic left ventricle (*LV*; atrioventricular discordance). The right ventricle gives rise to the aorta, and the left ventricle gives rise to the main pulmonary artery (transposition or ventriculoarterial discordance). The position of the ascending aorta (*A*) to the left of the main pulmonary artery (*PA*) indicates that this patient has levotransposition. Reprinted with permission from Reddy and Caputo [68]

the trigger point for manifestation of the condition. There are several case reports in the literature of late incidental diagnosis of ccTGA, with both underlying dextrocardia and levocardia [30–33].

The data gathered from a Mayo Clinic study demonstrated the survival curve for these patients to be consistent at a rate of about 1–2 % per year of follow-up. Consequently, the average 10 year survival of children diagnosed with AV discordance was 64 % [34]. The worst prognostic factor for this cohort of patients was existing tricuspid insufficiency (at the moment of diagnosis), which significantly worsened the prognosis, while underlying VSD with or without corresponding pulmonary stenosis showed little or no effect on survival.

Heart block accompanies a high proportion of cases, along with L-looping, ccTGA, and situs solitus. It has been shown that 3–5 % of children with this anomaly will have complete heart block at birth. An additional 20 % will develop spontaneous heart block at some point during life, with a traumatic event like surgery being a well-recognized stimulating factor [35, 36].

AV concordance is also frequently associated with intrinsic heart defects. Two groups have demonstrated that patients with concordant hearts and corresponding congenital defects can progress to biventricular non-compaction. This is usually the result of arrested endomyocardial development in patients with CHD. Friedman et al. [37] and Baskurt et al. [38] described cases of dextroversion associated with non-compacted cardiomyopathy in the left ventricle, while Grattan et al. [39] presented dextroversion patients with accompanying biventricular non-compacted cardiomyopathy.

In summary, although dextrocardia with situs solitus rarely, if ever suggests any visceral pathology, it is typically accompanied by complex cardiac malformations and disruption of the conduction system. However, these patients may still present with small or even absent heart pathology. If patients are asymptomatic, they are often left undiagnosed, and cardiac referral is delayed until adulthood, when patients start exhibiting manifestations of acquired heart disease.

Situs inversus

Dextrocardia with situs inversus is a condition in which the heart and other viscera are a mirror image of normal. In this condition, the morphology of the structures does not reflect their common location. The morphologic right heart which pumps venous blood is on the left anatomic side, and the morphologic left heart is on the right anatomic side. Dextrocardia with situs solitus is the most common variant of heart misplacement disorders.

Unlike dextrocardia with situs solitus, the underlying mechanism of situs inversus is incorrect situs formation, rather than heart malrotation. Hence, other organs are mirrored as well, including the liver and gallbladder, located on the left, and stomach and spleen, located on the right. This is otherwise called situs inversus totalis. However, in some situations, the heart can still be found in the left hemithorax (due to malrotation)—this is called situs inversus with levocardia.

Various heart anomalies can occur with situs inversus, but in general this condition tends to be asymptomatic and less associated with significant heart malformations, compared to other types of dextrocardia. Only 5–10 % of cases present with substantial heart pathology [25, 40]. Although most authors agree that patients with situs inversus demonstrate a low incidence of cardiac abnormality, some authors suggest that there is a significantly higher incidence of heart defects in this group compared to normal hearts [19, 20, 41, 42]. This may be partially explained by the

characteristics of the population studied, which mostly represents patients referred to tertiary care centers. The actual incidence of cardiac malformations seen with in situs inversus is probably somewhere in between.

Among the others, the most prevalent cardiac defects associated with situs inversus are VSD and transposition of the great vessels, both corrected and complete [12, 43]. Profound defects such as univentricular AV connection (UVAVC) and double-outlet right ventricles (DORV) were also reported in the literature to be associated with situs inversus, although these were considered to be rare findings. Studies showed that situs inversus hearts tend to be normal when concordant connections between the atria and ventricles are present, similar to hearts seen in situs solitus dextrocardia. If malformations still occur in concordant hearts, they are likely to be DORV or complete TGA [29]. Some authors reported total anomalous pulmonary venous connection (TAPVC) and persistent right-superior vena cava (bilateral SVC) to be associated with situs inversus [44, 45]. Markedly, in situs inversus with levocardia, the frequency of heart defects is higher, close to the frequency seen in dextrocardia with situs solitus. This is due to more profound discrepancies during heart development. However, isolated levocardia with situs invertus (ILSI) is a rare condition: only 249 cases have been reported in the literature. In ILSI, the presence of a normal, acyanotic heart is atypical, with only 6.4 % of patients reported to have an acyanotic heart [46].

Heart chamber concordance is a good predictor of normal heart anatomy in situs solitus. Hence, as in the case of situs solitus, the AV connections should be assessed in all cases of situs inversus dextrocardia. As a rule of thumb, discordant heart connections are less likely to be found under these circumstances. Garg et al. [12] showed in an analysis of 125 patients with dextrocardia that 73–74 % of patients with situs inversus had a concordant heart. This finding exceeds by far the rate seen in patients with situs solitus. However, in cases with existing comorbidities, the outcomes of AV connection discordance are similar to the outcomes seen in situs solitus discordant hearts. Again, discordance may be associated with corrected and uncorrected TGA. However, in situs inversus, the inversely located arteries will represent D-TGA during correction, and L-TGA during complete transposition. This occurs due to the mirrored position of the heart.

As mentioned above, the rare discordant connection seen in situs inversus is accompanied by heart defects. In addition, due to a misaligned heart structure, one might also consider underlying defects of the conduction system, similar to the defects seen in situs solitus. This, however, is quite unlikely in the situs inversus discordant heart, as the posterior AV node connects to the posterior conduction bundles on the septum. This is similar to what happens in a normal, concordant heart, implying normal AV conduction in these patients [47]. There are also several reports of sick sinus syndrome diagnosed in patients with mirror-image dextrocardia [48–50]. The authors reporting this hypothesize on a possible linkage between mirror-image dextrocardia and sick sinus syndrome; however, the reports are limited and more evidence is required. In summary, heart rhythm disturbance is a less likely finding in patients with situs inversus, compared to situs solitus [51].

Another important clinical implication of situs inversus is association with Kartagener syndrome. This is characterized by impaired mucociliarry clearance that predisposes individuals to airway infections, bronchiectasis, chronic sinusitis, recurrent otitis media, and male infertility. While half of all cases of Kartagener syndrome represent partial Kartagener with situs solitus viscera, there is no association with dextrocardia, except for situs inversus. Research shows that during childhood and adolescence, the disease produces little impact on overall health. Respiratory testing in patients usually reveals little deviation, if any, from normal parameters [52]. However, the disease starts to manifest and cause deterioration of respiratory function in the mid-twenties of most patients. Therefore, the condition is mostly an incidental finding, especially in children and young adults. A stressful or traumatic event like surgery may act as a trigger point and produce symptoms of the disease. Ellerman and Bisgard [53] reported that respiratory function tends to be significantly worse in previously undiagnosed adults, compared to patients who were diagnosed in childhood and received proper follow-up. It is worth mentioning that perioperative respiratory infections are common. Manifestations of the disease are not only related to ciliary immobility, but are also due to abnormal neutrophil chemotaxis [54].

Kidney function is an additional major concern. While there are not many studies reporting deterioration of kidney function with Kartagener syndrome, there are several case reports demonstrating severe glomerulonephritis, which serves as a part of the pathologic process [3, 55].

Another important consideration is the association of situs inversus with spinal dysraphism and other spinal malformations [56–61].

Situs ambiguous

A number of complex cardiac defects are found in the hearts of patients with situs ambiguous, or heterotaxy. Situs ambiguous is the most severe and disorganized visceral misalignment disorder among all of the dextrocardia variants. As mentioned above, there are two types of heterotaxy: right-sided (asplenia) and left-sided (polysplenia). Since the 1-year mortality for asplenia patients is greater than 85 %, related to profound heart pathology as well as

other intrinsic pathology, anesthesiologists rarely encounter these patients. Moreover, if these patients are seen in the operating room, it will most likely be for a palliative cardiac surgery. For most of these patients, full biventricular repair is inapplicable [62]. Most infants born with asplenia syndrome along with right heart isomerism have a number of cyanotic heart defects, among which obstruction of the pulmonary outflow tract and pulmonary atresia are seen in the overwhelming majority of patients. Incompletely formed atrial septum is seen in most cases of right heterotaxy, but it is also prevalent in the left heterotaxy group. Left-sided heterotaxy, or polysplenia, is a less severe disease than right-sided heterotaxy, with which 50 % of patients survive into adulthood. Left isomerism of the left atrial appendages is one of the major characteristics of left-sided heterotaxy. Furthermore, left-sided heterotaxy is more feasible to repair. Although it may also be associated with complex cyanotic heart disease, it is not as frequent as right-sided isomerism. It was previously reported, however, that only one-third of patients with left heart isomerism have a serious heart defect, while two-thirds of patients present with simpler forms of defect [63]. Association of left heart isomerism with interrupted inferior caval vein without any other significant heart defect is the most common presentation of the disease. These patients are likely to be relatively asymptomatic, and may not be diagnosed until adulthood in the absence of extracardiac anomalies [64]. In some cases, the inferior vena cava (IVC) is known to drain into a left SVC. However, anomalous pulmonary venous drainage is a more common feature of right isomerism. As mentioned earlier, conduction problems are often seen due to aberrant anatomy, with doubling of bundles in right isomerism, and hypoplastic sinus node in left isomerism. AV block is rarely seen in right isomerism, but occurs about 10 % of the time in left isomerism [64].

In summary, right isomerism is almost always recognized in infancy due to its universal association with severe cyanotic heart lesions. Left isomerism involves a wider range of underlying pathology, most of which is not severe and thus does not warrant prompt surgical intervention. For this reason, many patients with left isomerism are often left undiagnosed. Extracardiac congenital defects are associated with both types of isomerism. On top of splenic abnormalities, a high incidence of renal tract defects and biliary atresia has also been reported. It was also reported that Asians are more prone to have a heterotaxy syndrome (32 % more prevalent among Asians compared to Westerners [65]).

Diagnosis

Physical examination of the heart, such as percussion and palpation during routine exams, are vitally important initial diagnostic instruments, as these may help a physician to suspect heart malposition. X-ray, CT scan, ECHO, and magnetic resonance imaging (MRI) are all invaluable imaging modalities to confirm and to classify the diagnosis of dextrocardia. Typical X-ray images are shown in Fig. 3. CT scans are demonstrated in Fig. 4. Figure 5 demonstrates representative MRI views with dextrocardia. An electrocardiogram of a patient with dextrocardia is shown in Fig. 6. However, the differential diagnosis between various types of heart malposition presents a challenging task and requires clarification.

There are several different factors which must be considered when forming the diagnosis of dextrocardia and assigning a specific subtype. Diagnosing dextrocardia requires a systematic approach to look at all the different parts of the heart anatomy, and to analyze any structural anatomic defects which may be present. One of the more commonly used approaches is the segmental approach, which was first described over 40 years ago by van Praagh, and recently updated by Lapierre [15, 66]. This method has 3 key steps, which follow a classic venoarterial sequence analyzing the atria, followed by the ventricles, and then the great vessels [67]. The segmental approach can be applied to a number of different imaging modalities, including multidetector CT and cardiac MR imaging [68]. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) have also been described as useful imaging modalities to diagnose dextrocardia [69]. TEE is usually utilized in patients with a poor TTE window.

The first step in the segmental approach is to describe the visceroatrial positioning, and to define the visceroatrial situs as situs solitus, situs inversus, or situs ambiguous. This can be done by using chest X-ray to visualize the stomach, liver and spleen, and by using cross-sectional imaging to visualize the atrial situs. The situs of the atria and the viscera are the same in situs solitus and situs inversus [70]. Specifically, CT cross-sectional imaging can be used to view the morphology of the right atrium, which can be identified based on the presence of a triangular appendage with a large ostium. Similarly, the morphologic left atrium can be characterized through CT by the presence of a narrow ostium and a tubular appendage [68]. TTE can also be utilized to define the atrial situs, by detecting the location of the IVC, hepatic vein, and aorta. If the hepatic veins and IVC are located to the right of the thoracic spine, and the aorta is located to the left of the thoracic spine, situs solitus can be diagnosed, while situs inversus can be diagnosed if a mirror image of the previously described structures is present. Situs ambiguous, on the other hand, can be diagnosed if both the aorta and IVC are seen on the same side of the thoracic spine.

The next step is to analyze the ventricular orientation—this includes describing the AV connections, ventricular

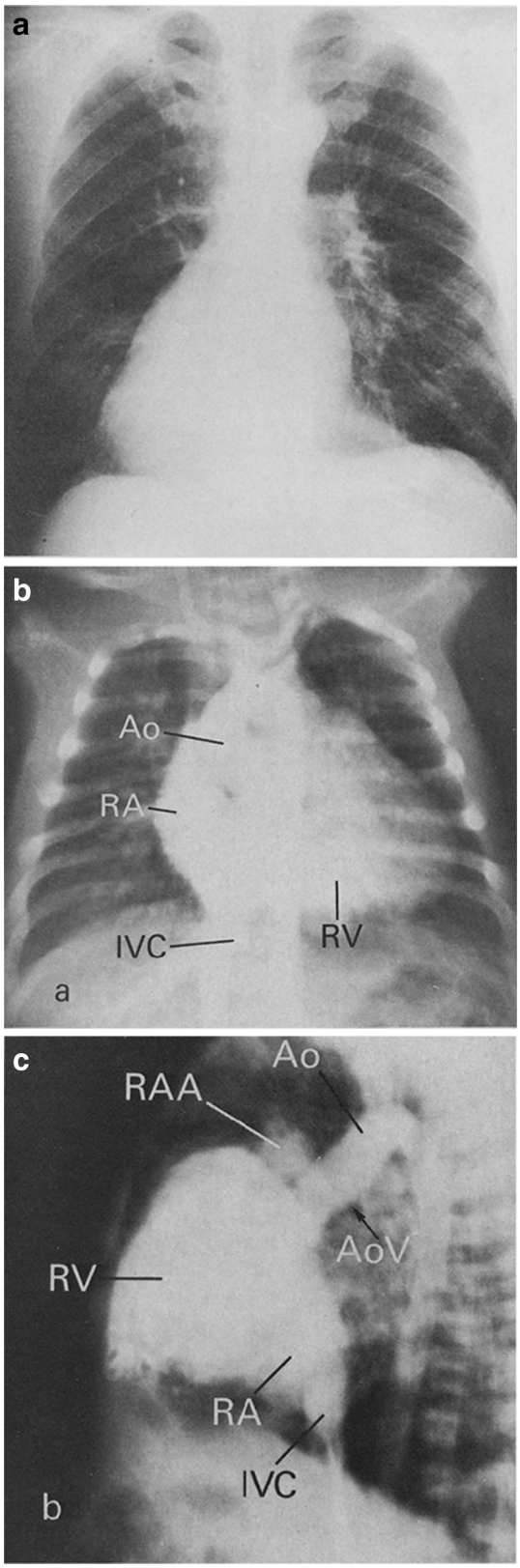

Fig. 3 **a** Posterior–anterior chest X-ray showing a left aortic arch, left descending aorta, and left stomach, but a right thoracic heart. Reprinted with permission from Buxton et al. [18]. **b** Dextrocardiogram following saphenous vein injection. Posterior anterior (**b**) and left lateral projections (**c**). The posterior and inferior location of the aortic valve is noteworthy in (**b**). *Ao* aorta, *AoV* aortic valve, *IVC* inferior vena cava, *RA* right atrium, *RAA* right atrial appendage, *RV* right ventricle. Reprinted with permission from Van Praagh et al. [88]

morphology, ventricular situs, chamber positions, ventriculoarterial connections, and the orientation of the great vessels, in this specific order [71]. This usually requires CT cross-sectional imaging, particularly when looking at the morphology of the ventricles [24]. However, the morphologic differences between left and right ventricle have also been identified using ECHO [12]. The AV connections are analyzed based on whether the right and left morphologic atria are connected to the right and left morphologic ventricles, respectively—this would be described as concordant. On the other hand, if the morphologic right atrium is connected to the morphologic left ventricle, and the morphologic left atrium is connected to the morphologic right ventricle, a discordant relationship would be assigned. The morphology of the ventricles is established based on the appearance of coarse trabeculae with moderator band (right ventricle) vs. fine trabeculae and a smooth septal surface (left ventricle). The morphologic right ventricle also has the infundibulum, a muscular portion of the outflow tract which separates the inflow valve from the outflow valve. Conversely, the left ventricle does not have an infundibulum, and thus there is a smoother, more gradual transition between the outflow and inflow valve tracts (mitral-aortic fibrous continuity) [26].

After completing the segmental approach, other malformations and anomalies must be noted and ruled out. Specifically, one must rule out anomalies associated with dextrocardia with situs soilitus, as well as situs ambiguous, as these malpositions are known to be associated with cardiac defects 90–100 % of the time. These cardiac defects include the presence/size of any atrial septal defect (ASD) and VSD, and the presence and level of any outflow tract stenosis. Types of great vessel hypoplasia and stenosis, including aortic coarctation and hypoplastic aortic arch, as well as the presence of patent ductus arteriosus (PDA), venous return, and other anomalies should also be noted. In addition, patients with situs inversus should be evaluated for potential signs of primary ciliary dyskinesia—these include those with a history of frequent respiratory infections and signs of hypoxygenation. CT may show lung overinflation, bronchial wall thickening, and sinus malformations. Electron microscopy may be utilized to analyze respiratory mucosa specimens. A decreased amount of nasal saccharine during exhalation may be detected during a simple breathing test. Furthermore, the cilia can be examined for structural defects, coordination, and beat abnormalities, and abnormalities of sperm motility can also be seen.

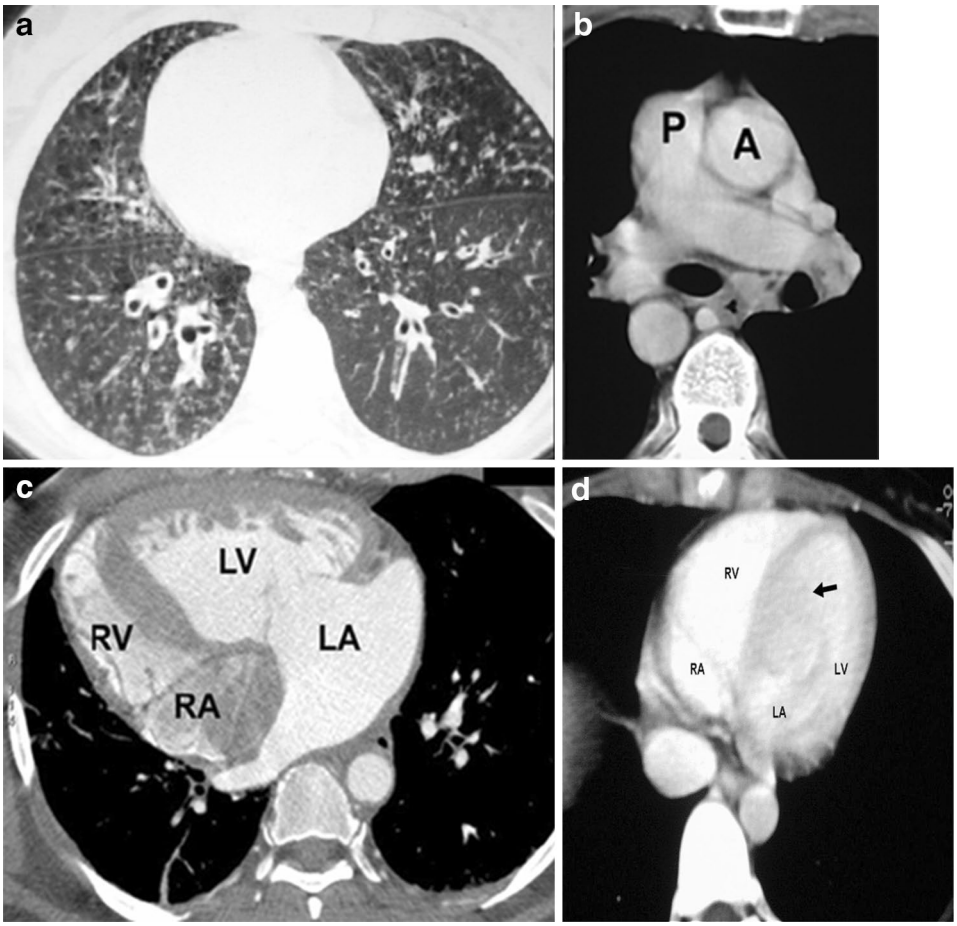

Fig. 4 CT scan in 22-year-old man with Kartagener syndrome (situs inversus). **a** Image of lungs shows dextrocardia and bronchiectasis. **b** Image of great vessels shows that main pulmonary artery (*P*) is to the right of ascending aorta (*A*), an inverted relationship, as is expected with situs inversus. **c** Axial image from ECG-gated CT scan at level of ventricles shows that the left atrium (*LA*) and left ventricle (*LV*) are anterior to right atrium (*RA*) and right ventricle (*RV*). Ventricles are in D-Loop configuration. **d** Oblique image in plane of outflow tract of left ventricle (*LV*) shows fibrous continuity between inflow mitral valve (*arrow*) and outflow aortic valve, confirming a morphologic left ventricle. Left ventricle is located anteriorly and inferiorly to right ventricle (*RV*). Reprinted with permission from Maldjian and Saric [24]

Clinical implications

In order to clearly describe the most important clinical implications of the various subtypes of dextrocardia, we have categorized the material in a system-based manner. Review of the material previously discussed, including knowledge of the natural history of dextrocardia, the embryologic basics of dextrocardia, and existing experiences in available case reports, will facilitate a better approach to managing dextrocardia patients. If dextrocardia is an incidental finding in a patient, then the patient should undergo the diagnostic algorithm in order to arrive at a correct diagnosis. Whether the patient has just been diagnosed or presents with a known history of heart malposition, there are important considerations of preoperative and intraoperative management.

Cardiovascular system

Because dextrocardia affects mostly the cardiovascular system, it is vitally important to rule out any underlying heart pathology, even if the patient is asymptomatic and was previously diagnosed with heart malposition. Patients with situs solitus dextrocardia, as well as patients with levo- or mesocardia, must be carefully assessed for atrial, ventricular, and arterial concordance, because these subtypes of dextrocardia suggest a very high incidence of underlying heart disorders. However, the number one concern, especially in asymptomatic patients with situs solitus dextrocardia, is congenitally corrected TGA. CcTGA should be suspected until proven otherwise, and the only indication of ccTGA may be an abnormal cardiac shadow location on an otherwise normal chest X-ray. However, associated heart defects should also be ruled out during thorough examination. Marked cardiomegaly, an increase in pulmonary vascular markings, and left atrial enlargement may raise increased suspicion of the presence of VCD or left-to-right shunt. Darkened lung fields on chest X-ray may suggest the presence of pulmonary stenosis. The routine preoperative assessment should always include electrocradiogram (ECG), chest radiograph, TTE, and pulse oximetry. TTE of a patient with dextrocardia is demonstrated in Fig. 7. If any pathology is suspected, further evaluation is necessary. In the event that coronary artery disease is found or coronary artery bypass graft is planned, coronary angiography should be performed. If ccTGA is detected by ECHO, full

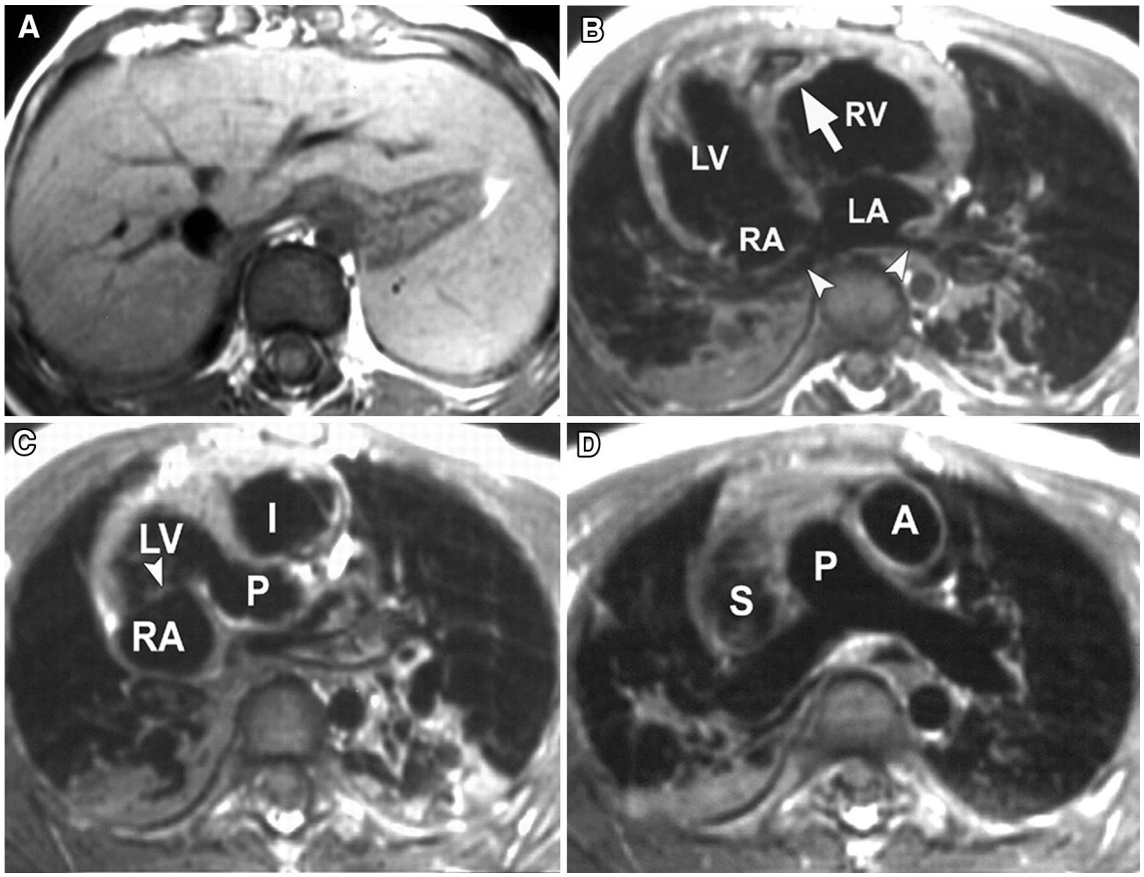

Fig. 5 ECG-gated axial spin-echo T1-weighted MR images in patient with dextrocardia, situs solitus, and corrected TGA. **a** Liver is on right and spleen is on left, revealing situs solitus. **b** Image through cardiac chambers shows discordant atrioventricular connections. Inferior pulmonary veins (*arrowheads*) drain to morphologic left atrium (*LA*). Left atrium is connected to morphologic right ventricle (*RV*), which is distinguished by presence of a moderator band (*arrow*). Morphologic right atrium (*RA*) is connected to morphologic left ventricle (*LV*). Ventricles are in L-loop configuration. **c** Images at progressively higher levels show muscular outflow tract or infundibulum (*I*), which is characteristic of a morphologic right ventricle. Pulmonary artery (*P*) arises from outflow tract of morphologic left ventricle (*LV*). Right-sided atrioventricular valve (*arrowhead*, top image) is near root of pulmonary artery because of fibrous continuity of inflow and outflow valves characteristic of a morphologic left ventricle. **d** Aortic root (*A*) arises from morphologic right ventricle. Reprinted with permission from Reddy and Caputo [68]

left and right heart catheterization is essential, regardless of the situs pattern. Catheterization can eliminate elevated ventricular end-diastolic pressures and intracardiac shunts, as well as tricuspid regurgitation [72]. If cesarean section is planned, it has an important implication in situs inversus. Aorto-caval compression is a common occurrence, because atypical location of the vena cava makes it problematic in deciding whether right or left displacement of the uterus is required [4].

Heart rhythm disturbances and ECG considerations

In order to correctly interpret electrocardiographic findings, the leads should be adjusted. For each particular type of dextrocardia, the electric potentials reflect differently, depending on the precise stereotypic position and situs type, because it is the situs that determines the course of the P wave [73]. In situs solitus, atrial depolarization progresses normally regardless of whether the location of the heart is normal or malpositioned. However, ventricular depolarization is counterclockwise. Q waves will appear in leads I and aVL. Q waves are also normally seen in the left precordial leads due to appropriate septal depolarization. Furthermore, K waves can be seen in leads V1 and V2, with RS complexes in the remaining chest leads [74]. Hence, for situs solitus and isolated dextrocardia, the chest leads should be reversed, while the limb leads should remain unaffected [75]. This results in corrected limb leads, as the situs type is normal, and the heart axis points to the right.

In contrast, in the case of situs inversus, the sinus node is located to the left (mirrored), so the P wave is now inverted. This means that there is a negative P wave in leads I and aVl, and a positive P wave in aVR. Situs inversus is also associated with inverted ventricular activation,

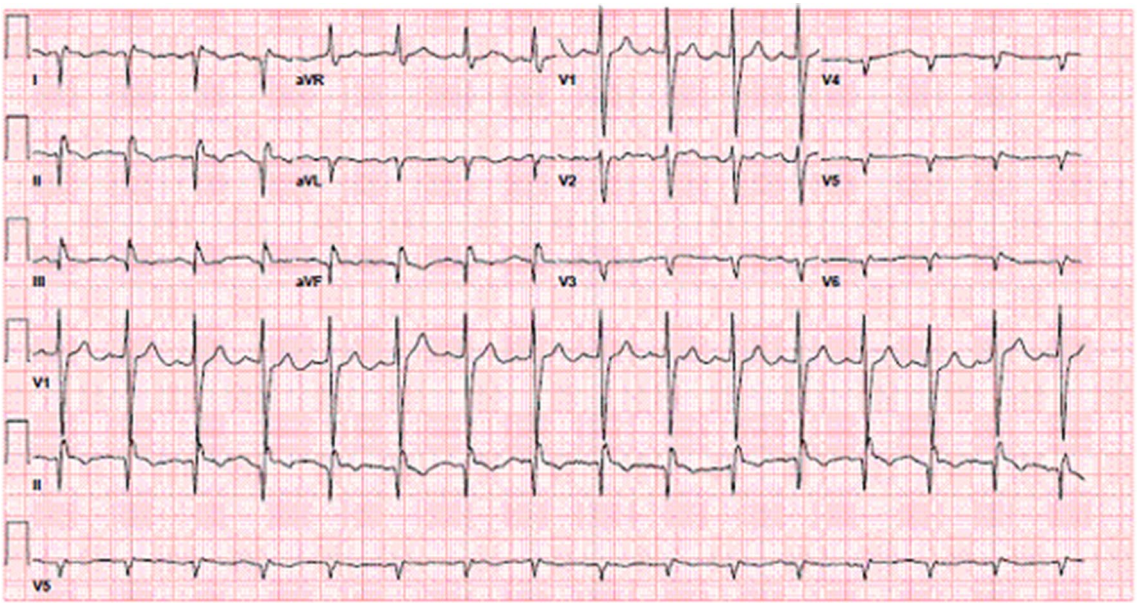

Fig. 6 ECG (normal lead positions) showing marked right-axis deviation of the P wave (negative in aVL and lead I) and of the QRS complex, and low voltage in the precordial leads, V4 through V6. Lead aVR is similar to the normal aVL in the normal ECG. Reprinted with permission from Bindra et al. [89]

and reversed repolarization. It is no surprise that the QRS complex is negative in lead I, while the T wave is inverted. Lead aVR resembles aVL and vice versa, while the right precordial leads mirror the left precordial leads in a normal heart. Also, mirror positioning of the heart results in right-to-left septal depolarization, so Q waves will be present in the right precordial leads. In order to obtain correct readings, the limb leads should be reversed, and the chest leads also inverted and recorded from the right precordium.

In situs ambiguous and right isomerism, different origins of P waves may be present at different times—these represent the activity of the bilateral sinus nodes. However, the P wave axis can still be normal if the right sinus node acts as a dominant pacemaker. Due to the absence of a functional sinus node, patients with left isomerism have an ectopic atrial pacemaker, and the P wave is usually abnormal. As an ectopic beat, the location is different, and there is thus slower firing. Because of this, patients experience progressive slowing of heart rate with age. Most patients tend to require placement of a permanent pacemaker [76]. Conduction of the ectopic atrial pacemaker is frequently blocked, which leads to nodoventricular incoherence and a narrow QRS. Complete heart block is a major cause of morbidity and mortality.

Cardiopulmonary resuscitation (CPR)

In the case of life-threatening heart arrhythmias resulting in cardiac arrest, cardioversion needs to be applied with the defibrillator paddle placed on the right side of the chest. While CPR guidelines state that paddles may be placed in several different positions to be equally effective in treating ventricular or supraventricular arrhythmias (anteroposterior, anterolateral, anterior–right and anterior–left infrascapular), left anterolateral positioning is futile in the case of dextrocardia, as the heart's electrical axis is mirrored, and thus no electrical stimulus reaches the heart. Right anterolateral positioning, however, is more effective [77]. Furthermore, chest compressions should be applied slightly to the right of the chest, to effectively compress the heart.

Vascular access

The mirrored position of the thoracic viscera (situs inversus) has an impact on common anatomic landmarks. This is especially important when placing a central line. Typically, a correctly positioned right subclavian cannula remains on the right side, without crossing the midline. However, in the case of situs inversus, correct cannulation presumes crossing the midline. Moreover, total anomalous pulmonary venous connection (TAPVC) and right superior vena cava (SVC) are reported to accompany situs inversus and situs ambiguous. If the subclavian vein cannula does not show up on the opposite side of placement, entry into the right SVC, TAPVC, or malposition into the aorta should be suspected. In these cases, a right internal jugular vein (IJV) is a preferable site for central venous cannulation (CVC), because it usually flows directly into the superior vena cava [45]. The right side is the generally preferred CVC insertion

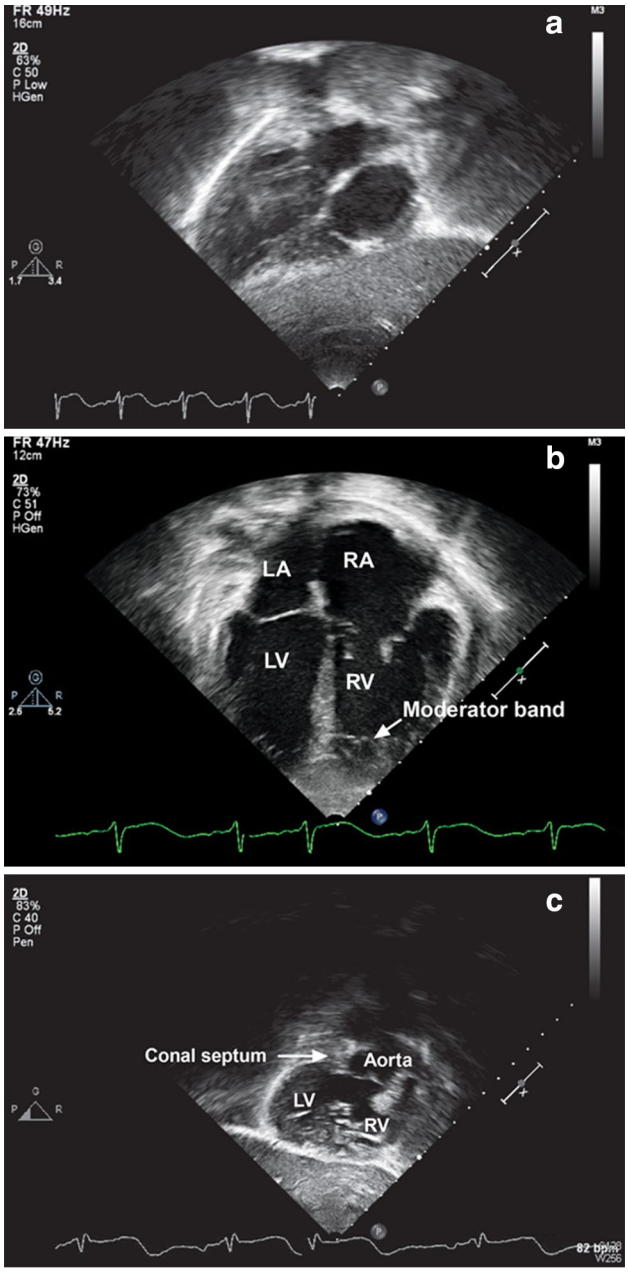

Fig. 7 **a** Tetralogy of Fallot associated with dextrocardia and situs inversusa. Transthoracic echocardiogram (subxiphoid long-axis view) shows a rightward cardiac apex. **b** Transthoracic echocardiogram (apical 4-chamber view) shows dextrocardia and situs inversus. *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle. **c** Transthoracic echocardiogram (subxiphoid long-axis view) shows the anteriorly deviated conal septum causing subpulmonic stenosis: a large cono-ventricular septal defect with severe right ventricular outflow tract obstruction. Reprinted with permission from Dilorenzo et al. [90]

site in cases of situs inversus as well, due to the fact that the thoracic duct is on the left, and also because the right lung is slightly lower than the left lung [46]. Rightward insertion may lower the risk of perforation of these structures.

Central venous cannulation, as well as brachial plexus blockade, may pose additional challenges with dextrocardia, especially when associated with situs inversus. Ultrasound guidance should be utilized, if possible. It is advisable to use ultrasound guidance during arterial cannulation and pulmonary artery catheter insertion as well. Due to an unpredictable vascular course, injecting a small amount of air with saline into the central line may be helpful in identifying proper subclavian or internal jugular veins draining into the atrium after cannulation [77]. It is also preferred that cannulation be performed on the left side [3].

If surgery requires a cardiac bypass, the location of the aortic arch should be ascertained before cannulation, because the aortic arch could be right-sided [21]. In total situs inversus, the aorta is usually posterior and to the left of the pulmonary trunk [78]. As mentioned, anomalous venous drainage and persistent right SVC have been reported with situs inversus, so should bicaval access be required, bilateral SVC may impact the approach, and should be ruled out prior to surgery [79]. It should also be noted that in dextrocardia with situs solitus, the right atrial appendage is usually difficult to reach.

Respiratory system

Isolated dextrocardia (situs solitus) rarely includes any underlying airway pathology, and can be managed routinely. In contrast, situs inversus is associated with primary ciliary dyskinesia (PCD or Kartagener syndrome) in 25 % of cases. Airway mucous retention and an inability to clear pathogens are the primary abnormalities in Kartagener syndrome, as there is an impairment of mucociliary clearance. These patients are usually encountered during pulmonary surgery for bronchiectasis, or ENT surgery for sinusitis or otitis media. In both cases, respiratory complications are the number one concern that the anesthesiologist will face in this patient group. Antibiotic prophylaxis, administration of bronchodilators with chest physiotherapy, postural drainage, and incentive spirometry are all required. Using disposable air equipment and performing smooth manipulations during intubation and extubation are required steps to avoid traumatizing the mucosa. The patient's pulmonary status should be optimized prior to the surgical procedure if general anesthesia is planned. Bronchoconstriction can be avoided by using volatile anesthetics, as they directly relax the bronchial smooth muscles [80]. Patients with pre-existing spastic bronchial disorders combined with coronary pathology will benefit from pretreatment with α_2-adrenergic agonists. In order to prolong the expiratory time, the respiratory rate should be kept low [81]. A heated humidifier or moist heat exchanger should be used at all times.

Pulmonary inversion is another factor that needs to be considered, as left main stem intubation is possible. When

one-lung ventilation is planned, the inversion should be considered when inserting the double-lumen tube. There have been cases of life-threatening intraoperative bronchial obstruction complicated by subsequent postoperative pneumonia, despite having undertaken all preventive measures [82]. Postextubational mini-tracheostomy may be required for the evacuation of residual secretions. It is recommended to obtain informed consent for possible mini-tracheostomy prior to surgery from every patient with Kartagener syndrome.

Otherwise, a regional block is the method of choice, provided that the motor block does not spread high into the thorax and respiratory muscle tone is maintained, allowing for spontaneous breathing. Not only does the block help to prevent respiratory complications intraoperatively, it also allows for early postoperative ambulation and contributes to improved clearance of respiratory secretions. However, certain spinal deformities such as spina bifida, meningomyelocele, and split cord all reported with Kartagener syndrome may prevent the use of neuraxial anesthesia. Patients should be evaluated carefully prior to surgery, especially pediatric patients, as they are more likely to present with hidden abnormalities.

Masseter spasm and difficult intubation due to anterior laryngeal position, as well as prolonged post-anesthetic apneic period (a duration of 30 min) have been reported in association with situs inversus and Kartagener syndrome [83]. A case of prolonged paralysis after administration of succinylcholine showed a possible link between situs inversus and atypical cholinesterase [83, 84], although no other similar reports exist in the literature, so routine testing for atypical cholinesterase is not recommended at this point.

Immune system

Defective chemotaxis has been observed in patients with Kartagener syndrome. The abnormalities are related to impaired motility of neutrophils [54], and thus aseptic techniques during anesthesia are of paramount importance; this includes during placement of a needle and catheter insertion for epidural block. Heinz or Howell–Jolly Bodies found on a peripheral blood smear of a patient with heart malposition indicate that asplenia is present, and that there is a decreased immunity to encapsulated bacteria. Antibiotic prophylaxis is generally recommended for all patients with right heterotaxy syndrome as well as for any other patients with congenital asplenia. Specifically, Dyke et al. [85] reported that patients with asplenia syndrome are at an increased risk of developing fatal septicemia.

Renal system

It should be noted that there is some association between situs inversus and renal abnormalities. Polycystic kidney disease and glomerulopathies can depress kidney function and influence renal metabolism. Thus, kidney function should be tested preoperatively if situs inversus with Kartagener syndrome is suspected.

Summary of clinical considerations

Perioperative clinical considerations primarily include preoperative assessment and intraoperative management. An outline of a basic algorithm for appropriate management of the patient with dextrocardia is provided in Fig. 8.

Preoperative

1. If the patient has a diagnosis of Kartagener syndrome, certain preoperative pulmonary issues should be identified or addressed before proceeding with surgery. These patients are predisposed to multiple airway anomalies and infections that can cause significant perioperative issues. The presence of $PaCO_2 > 50$ mmHG may signal the possible need for postoperative mechanical ventilation. Pulmonary percussive therapy, bronchodilators, incentive spirometry, postural drainage, steroids, and antibiotics are necessary to optimize the pulmonary system before surgery. Premedications which cause hypoventilation or depress mucociliary function should be avoided if possible in these patients [86].
2. Dextrocardia, alone, is associated with multiple congenital heart anomalies, so anatomic cardiac knowledge should be acquired before surgery. Patient records of prior cardiac procedures and work-ups should be ascertained. Patients with congenital heart disease frequently consider surgery curative and considered themselves “fixed”. Only patients with certain ASDs and patent ductus arteriosus are cured after surgery. The remaining congenital heart patients have residual issues which require evaluation and possible intervention before progressing to noncardiac surgery.

Intraoperative

1. Patients with situs inversus require certain intraoperative considerations. The ECG and pacing or defibrillator pads should be placed in reverse. If not, the polarity change for the ECG can erroneously display a picture of perioperative ischemia. Successful cardiopulmonary resuscitation and defibrillation requires proper knowledge of cardiac anatomy. Transesophageal echocardiography imaging and interpretation must take into account the possibility of uncovering abnormalities missed in previous work-ups. Central venous cannulation should occur in the left internal jugular vein,

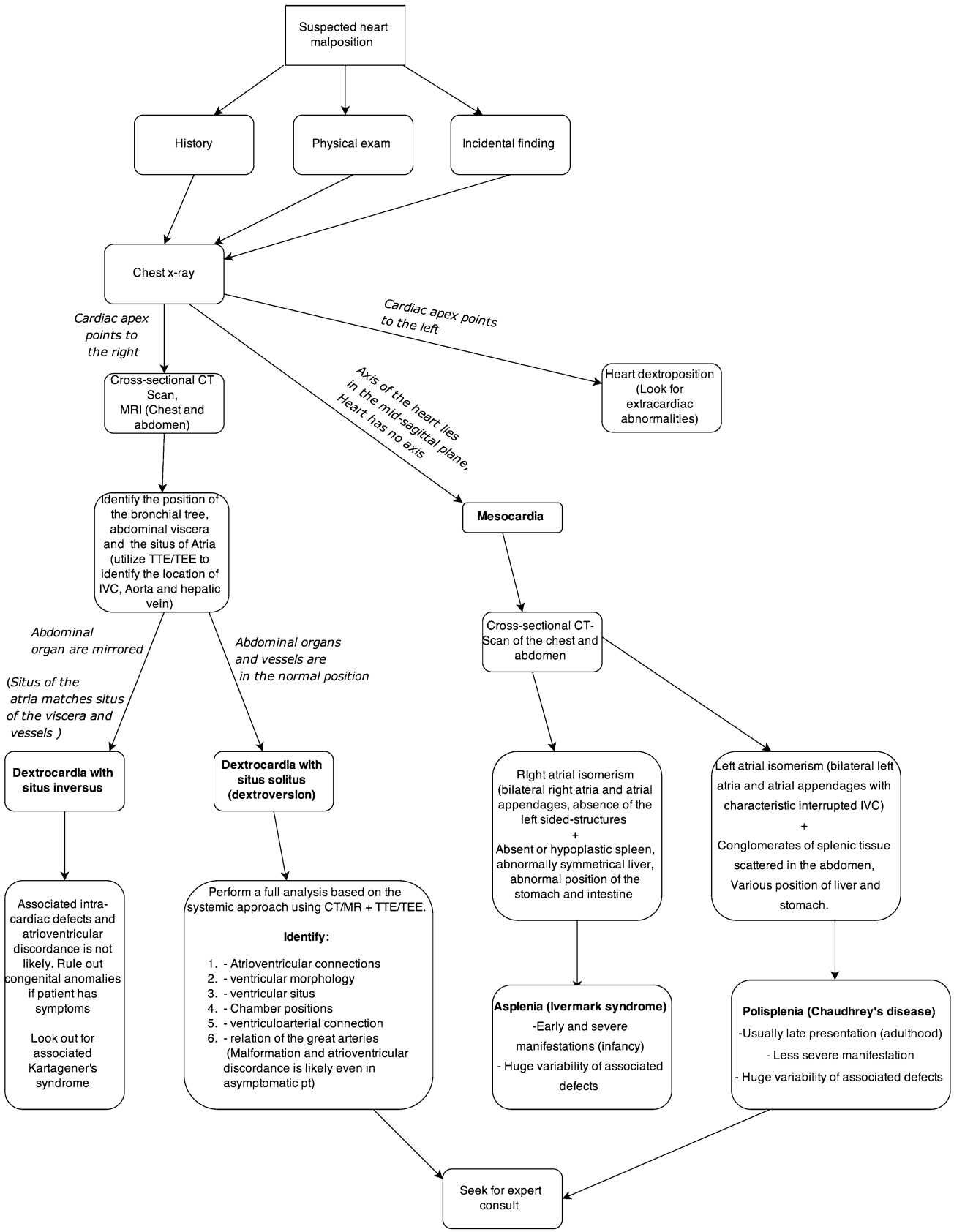

Fig. 8 Algorithm of appropriate management of the patient with dextrocardia

which provides a direct route to the right atrium and lessens the incidence of thoracic duct injury. The left mainstem is usually entered by an endotracheal tube travelling past the carina. Finally, in the obstetric arena, uterine displacement should be to the right.

2. In patients with Kartagener syndrome humidification of gases should be added to the anesthesia circuit and frequent suctioning of the endotracheal tube may be required. Placement of a double lumen endotracheal tube will require fiberoptic guidance in the light of possible anatomic variations and heavy mucoserous secretions encountered by the anesthesiologist. Nasal airways and/or nasal intubation should be avoided in these patients due to the high incidence of sinusitis. Regional anesthesia is preferred for these patients if possible [87].

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