Management strategies for interrupted aortic arch with associated anomalies

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Summary

Interrupted aortic arch (IAA) is characterised by a lack of luminal continuity between the ascending and descending thoracic aorta. It represents a critical ductus dependent congenital heart disease, which without surgery is associated with high mortality in the neonatal period. Management remains challenging, although in recent years overall mortality appears to be improving. Long-term morbidity and need for repeated interventions are areas of concern. It is difficult to make generalised recommendations based on this literature review as complex associated anomalies often require individualised management strategy.

Keywords: Congenital heart disease; Aorta; Interrupted aortic arch; Associated anomalies

1. Introduction

Interrupted aortic arch (IAA) was described by Steidele in 1778 and is now defined as an uncommon congenital cardiovascular malformation characterised by the lack of luminal continuity between the ascending and descending thoracic aorta [1,2]. Untreated, the median age at death was 4—10 days, usually following physiological closure of the ductus arteriosus [2,3].

Factors contributing to improved results in recent years include accumulated surgical experience, the care of patients with congenital heart diseases centralised into major centres, antenatal diagnosis, better stabilisation and treatment of all complications before definitive repair [4,5]. Although operative survival has improved in recent years, further interventions are often required to address residual or recurrent left heart outflow or residual arch obstructions [4,5].

2. Incidence

IAA with ventricular septal defect (VSD) is an uncommon lesion, accounting for 1.5% of all congenital heart disease [6]. Most reports highlight type B morphology as the most common form (52—90%) [4,5,7,8]. A higher incidence of type A morphology (62.5%) could be found in patients with associated truncus arteriosus (TA) [4,9]. However, in some series type B was more common even in presence of TA [10,11].

3. Morphology

IAA is defined as either a complete discontinuity or a nonpatent fibrous strand in the transverse arch or aortic isthmus [12,13]. The types of IAA were described by Celoria and Patten [12,14]. They classified this lesion into three types, based on the site of aortic arch interruption. In type A, the interruption is distal to the left subclavian artery. In the most common type B, the interruption is between the left common carotid and left subclavian artery. In type C, which is the rarest type, the interruption is between the innominate and left carotid arteries.

There was generally a persistent arterial duct supplying blood to the lower body. Most patients have only an isolated VSD [13] but IAA can be associated with a variety of complex cardiovascular anomalies [1,7]. A spectrum of left ventricular outflow tract obstructive (LVOTO) lesions, particularly subaortic stenosis (SAS), is also common [1,4,5,7,15]. The malalignment of the conal septum with the resulting SAS is thought to be responsible for the in utero involution of the ascending aorta and aortic arch (fourth aortic arch) and there is reciprocal development of the pulmonary artery and ductus arteriosus (sixth aortic arch) [6]. The degree of SAS therefore is directly proportional to the size of the pulmonary
trunk and inversely proportional to the size of the ascending aorta [6,15].

4. Associated anomalies

Associated heart lesions are present in many patients: multiple VSDs in 2.7%, TA in 10–18% [6,10,16,17], LVOTO in 10–16%, valvular aortic stenosis (AS) in 10.5%, transposition of great arteries (TGA) in 4–6%, double outlet right ventricle (DORV) in 5.3% and aorto-pulmonary window (APW) in 5.3% [4,5,9,18]. Some authors reported much lower (3–4%) incidence of TA [12,19,20].

5. DiGeorge syndrome and deletion

According to Van Mierop about 68% of patients with IAA and about 33% patients with TA have DiGeorge syndrome [21]. It is typically associated with the type B IAA [4,7], which could explain the lower incidence of DiGeorge syndrome reported in some series as the incidence of type B was lower in these series [4,6]. However, the whole concept of DiGeorge and its testing was not well established when early series were reported [13].

Complete DiGeorge syndrome is defined as the coexistence of IAA with dysmorphic facial features; hypocalcaemia and low (<400 cells/mm) CD4+ T-lymphocyte count [3,6]. Partial DiGeorge syndrome is defined as the coexistence of the dysmorphic facial features with hypocalcaemia and IAA in the absence of any deficit of the CD4+ T-lymphocyte subpopulation [6,22,23].

Chromosome 22q11 deletion is associated with anomalies of the aortic arch in 24% of cases [24–26]. Guidelines are being developed for genetic screening of patients with aortic arch anomalies [24,25].

6. Diagnosis

A correct prenatal diagnosis of IAA and its different types is possible based on echocardiographic examination [26]. Thymic hypoplasia or aplasia may be diagnosed during fetal echocardiography that allows identification of a group at high risk for 22q11 deletion [27].

Gadolinium-enhanced 3D MRA (magnetic resonance angiography) may provide a detailed imaging of IAA that may not be possible with other non-invasive modalities [28,29].

A correct prenatal diagnosis should enable clinicians to provide the parents with further informative counselling and to plan adequate post-natal medical interventions [26,30].

7. Management strategy

IAA represents a ductus dependent congenital heart disease, which without surgery is associated with high mortality in the neonatal period. Recent advances in neonatal cardiac surgery have made it feasible to repair IAA-VSD within the first month after birth [6–8]. During the last several years, results of both primary and two-stage repair of IAA have improved, and early mortality in some centres approaches 8–10% [7,31–33]. However, the long-term fate of early survivors remains uncertain [4,5,34]. There is a risk of development of restenosis at the site of the aortic anastomosis and LVOTO [4,5,34–36]. Other residual lesions and non-cardiac complications are also described [4,5,33,34]. These often require reintervention and represent a risk for late death [4,5,14,33,34,37].

7.1. Preoperative resuscitation

Preoperative management includes prostaglandin, and may include assisted mechanical ventilation and dopamine infusion [7,32]. The aim is to improve lower body perfusion, improve renal function, diminish acidosis, and maximise the pulmonary/systemic resistance ratio across the ductus arteriosus [7]. A high index of suspicion is needed for necrotising enterocolitis [32]. Cardiac catheterisation should be avoided and MRI scan considered if echo is inconclusive [7].

7.2. Age at repair

In earlier series mean age of repair was 40–45 days [19,20]. Currently, the optimal time to operate is within first week of life due to pulmonary overcirculation [7,12,13,15].

7.3. Surgical repair

Successful surgical repair of IAA was first accomplished by Samson et al. in 1955 by direct anastomosis of type A IAA [1,38]. The associated VSD was not closed at the time of the arch repair [38]. Barratt-Boyes et al. in 1970 repaired type A IAA using a synthetic conduit [1,39]. In 1975 repair of a type B IAA with a direct anastomosis and closure of the VSD was reported [1,40].

Resection of all ductal tissues and extensive mobilisation of the descending aorta is important to prevent restenosis [7,32]. The techniques employed to repair the aortic arch are direct anastomosis between ascending and descending aorta; direct anastomosis with patch augmentation or a conduit interposition [7]. Some authors would recommend absorbable sutures to prevent restenosis [35]. In cases with contraindications to CPB and full heparinisation (e.g. intracerebral bleeding) aortic arch continuity can be restored through a left thoracotomy without use of CPB [7].

The VSD can be closed either through the pulmonary valve or right atrium depending on its location [7].

7.3.1. Single stage versus staged repairs

There are advantages to one stage repair over the staged approach which included fewer reoperations/reinterventions, avoidance of pulmonary artery banding (PAB) which could accelerate subaortic stenosis and the decreased need for future arch reconstruction [1,4,5,9,18]. Brown et al. on the other hand believed that pulmonary banding used in staged repair did not increase the development of subaortic stenosis and may in fact decrease subaortic stenosis [1].
A two-stage approach has been recommended in premature neonates weighing less than 1500 g, in the presence of severe infection, intracranial haemorrhage, multiple organ failure and very unfavourable morphology [9,33]. Several other authors have recommended a two-stage repair in the presence of complex anatomies like multiple VSDs, transposition of the great arteries, single ventricle [7]. However, some authors recommended a primary repair protocol in patients with low body weight [41]. In the group of patients after primary repair the early postoperative course often required delayed chest closure, inotropic support and prolonged ventilation [4,5,9,42].

Complete primary repair has obvious appeal, but the complexities encountered in achieving this goal could be associated with a higher mortality [1,7,8]. Recently, some authors reported a lower mortality with primary repair [4,5,9,18], but others reported a reduction in mortality with staged approach (31% to 19%) [1]. Serraf et al. reported no statistical difference in the mortality rate between single-stage and two-stage repairs [7].

Currently a single-stage biventricular repair of aortic arch obstruction and associated intracardiac defects seem to be the preferred technique at most institutions [1,4—6,9,18,20,43].

7.3.2. Techniques for repair of the aortic arch

A direct anastomosis after adequate mobilisation of both ascending and descending aorta without patch augmentation is thought to provide satisfactory patency and growth [1,7,31,36,44]. However, it carries the risk of recurrent obstruction due to tension at the anastomosis site or inadequate resection of ductal tissues [31,32]. It can also result in bronchial compression [31]. The distance between descending and ascending aorta could be as long as 2—3 cm in some patients with type B IAA, especially in the presence of aberrant right subclavian artery, and when the ascending aorta was extremely narrow and shifted more to the right [4]. In these cases it may be necessary to transect one or both subclavian arteries to increase aortic mobility [4,32]. Use of prosthetic conduit to restore continuity is considered suboptimal due to lack of growth potential [3,31,32].

Direct anastomosis with patch augmentation is considered by many surgeons to be an optimal method of repair [4,5,13,31,43]. A homograft [1,43] or autologous pericardial patch [1,7,8] can achieve a tension-free anastomosis with low incidence of recoarctation [4,5,13,31,43,45].

A left carotid artery swing down as an autologous conduit for arch reconstruction is an option to avoid recurrent arch obstruction and left bronchial compression [1]. It can be combined with a reverse subclavian flap to augment the diameter. In one report, sacrifice of the left carotid artery did not result in any adverse neurologic sequelae or growth disturbances [1].

7.3.3. Subaortic stenosis (SAS)

Patients with IAA often have posterior deviation of the conal septum, which morphologically can cause SAS and even aortic annular hypoplasia leading to varying degrees of LVOTO [1]. It presents a complex surgical problem [6,7,46].

The presence of LVOTO may influence both early and late survival and account for many late interventions and deaths [7,8,13,14,32—36,47,48]. Obstruction can be at multiple levels such as posterior malalignment of the conal septum, aortic valve hypoplasia or stenosis, and ascending aorta hypoplasia [32]. Predisposing risk factors are lower birth weight, type B IAA, single ventricle, bicuspid aortic valve, or anomalous right subclavian artery [13].

7.3.3.1. Diagnosis of SAS. There is a lack of uniform diagnostic criteria [6]. The presence of a nonrestrictive VSD makes use of physiologic parameters (e.g. preoperative subaortic pressure gradient) unreliable [6,7]. Hence, anatomic measurements must be employed [6]. When the subaortic diameter is smaller than two-thirds of the aortic annular diameter, SAS is considered to be severe [7]. The estimates of the subaortic diameters should be drawn from the level of maximal narrowing [6]. The systolic and diastolic diameters of the subaortic region, normalised to the diameter of the descending thoracic aorta at the diaphragmatic hiatus can be another parameter [6,11,49]. It is suggested that systolic ratios less than 0.6 and diastolic ratios less than 1.0 are indicative of severe SAS and mandate relief of the obstruction [50—52].

7.3.3.2. Relief of SAS. There is no consistency regarding which structures or measurements most optimally predict whether LVOT obstruction needs to be addressed at the time of an arch repair [13]. Jonas et al. do not recommend any intervention unless the subaortic stenosis is extreme, which means the diameter of the LVOT was less than about 3.5 mm in a neonate [4,5,8,9,53]. Others recommend a conservative approach when the LVOT diameter is greater than the baby’s weight +2 mm and a LVOT bypass procedure (Norwood) if the LVOT diameter was less than the baby’s weight in millimetres [54]. If the LVOT diameter falls in between, no definitive recommendation can be made [54].

Several procedures have been proposed to address subaortic obstruction [6,7,20,55—57]: (1) standard VSD closure ignoring the SAS [6,7], (2) creation of a double outlet left ventricle associated with aortopulmonary anastomosis and conduit insertion between the right ventricle and pulmonary artery bifurcation [19,20], (3) conversion into a univentricular physiology [7], (4) resection of the conal septum [7,55] and (5) application of the patch to the left aspect of the VSD (conal septum) [6].

Staged or primary repair without concomitant procedure to alleviate the SAS is identified in some reports as a risk factor for early death [6,8]. Staged approaches at repair of this complex lesion by reconstructing the arch and pulmonary artery banding proved unsuccessful [6,56]. Procedures that used extracardiac conduits to bypass the level of LVOT obstruction [6,20,56—59] can potentially achieve a biventricular repair at the cost of considerable early mortality and need for technically difficult reoperations [20,56—59]. Similarly conversion to univentricular physiology by means of a modified Norwood procedure is associated with significant mortality, morbidity and need for multiple reoperations and should be considered only in extreme cases (e.g. coexisting aortic valve atresia) [8,57].

LVOT obstruction can be relieved with resection of the posteriorly displaced conal septum [6,55,56,60,61]. Transaortic resection of the conal septum is technically difficult
because of the small size of the ascending aorta and aortic valve, and there is a high risk of aortic valve injury [67,55,56]. A transventricular approach is technically easier [67,55,56,60]. However, infundibulotomy (on RVOT) on the neonatal heart could have detrimental haemodynamic consequences [6]. Transatrial resection was associated with a significant occurrence of complete heart block (16%) [6,55,60], intraoperative aortic valve injury (17%) [55] and other postoperative complications.

Luciani et al. emphasised that adequate relief of the SAS cannot be achieved without re-establishment of a laminar flow through the LVOT [6]. They described a simple technique for relief of severe subaortic stenosis without resection of the conal septum [6]. A technically easier transpulmonary approach is proposed to expose the VSD and subaortic region [6]. The patch for closure of the VSD is applied on the left side of the septum to pull the conal septum into the right ventricle [6,7]. Placing the apical VSD patch stitches on the left ventricular side of the crista supraventricularis and downsizing the patch, can help the conal septum get deflected away from the subaortic region [6,7]. The oblique orientation of the VSD patch in this technique is critical in keeping the spur of conal septal tissue away from the subaortic area, recreating a smooth-surfaced LVOT [6]. This approach is reported to have statistically significant advantages over other approaches for SAS [6,7]. Luciani et al. [6] reported better survival, absence of procedural complications (heart block, valvular insufficiency) and adequate growth of the subaortic region with no recurrence of significant subaortic obstruction.

7.3.4. IAA with complex malformations

Association of complex malformations presents a surgical challenge [7,10,62,63]. Palliation often gives unsatisfactory results and complete repair should be undertaken at birth [7]. This is particularly the case for patients with truncus arteriosus, aortopulmonary window and transposition of great arteries [7].

7.3.4.1. Truncus arteriosus (TA). Only few large series of operated patients with TA and IAA were found in the literature [4,10,11,16,17,64–66]. The optimal timing of surgery, methods of heart and brain protection, aortic arch repair, truncal valve repair as well as the right ventricular to pulmonary artery continuity reconstruction continue to be debated [4,10,11,16,17]. The association of truncus arteriosus with IAA is usually free of LVOT obstruction [13]. However, presence of severe truncal valve insufficiency and coronary artery anomalies are risk factors for death [66].

7.3.4.2. Single ventricle. Patients with single-ventricle physiology and aortic obstruction usually have a large pulmonary artery and excessive flow [67]. Initial palliation with PAB (preservation of the pulmonary vascular bed) and repair of aortic obstruction, is associated with very low mortality [67,68]. It avoids brain ischaemia, cardiopulmonary bypass, and the associated morbidity [67]. This strategy can maintain acceptable anatomy and haemodynamics for later bidirectional Glenn and Fontan procedures [67,68]. However, PAB is implicated in the accelerated development of subaortic obstruction that ultimately jeopardised Fontan candidacy [67–76]. Other complications associated with a poorly positioned band or migrations are reported, including branch pulmonary stenoses, semilunar valve insufficiency, and erosion with aneurysm formation [67,69]. These deleterious effects prompted surgical strategies seeking early initial relief of aortic obstruction with avoidance of PAB [69–76]. A palliative arterial switch operation is advocated for such newborns trading subaortic for neosubpulmonary stenosis [67,77,78]. It may lead to PA distortion complicating subsequent Fontan. A Norwood procedure is an alternative option for primary palliation, as it provides relief at all levels of systemic obstruction along with a source of pulmonary blood flow [71,72,77]. In patients with arch obstruction and an ascending aorta smaller than about 4 mm in diameter, the arch can be repaired with a DKS type of repair [72]. Disadvantages with the DKS procedure, especially in the presence of arch obstruction, are high early mortality [72,79,80], development of semilunar valvular dysfunction [72,80–82] and usually a requirement for deep hypothermic circulatory arrest, with risk of neurologic insult [72,83,84].

A consensus seems to be emerging that patients with single-ventricle physiology and systemic obstruction should undergo aggressive neonatal surgical relief from all potential residual systemic obstruction by the Norwood procedure [7,61].

7.4. Reinterventions

The long-term probability for reoperation or reintervention remains regardless of the operative technique chosen [3,5,7,21,23,26]. A high rate of development of restenosis at the site of aortic anastomosis, LVOTO and left bronchial compression has been reported [1,3,4,12,13]. Some authors report a higher occurrence (25%) of LVOTO after primary repair [4,5].

7.4.1. Aortic arch stenosis

Fulton et al. [47] noted a freedom from reintervention for arch obstruction of 83% at 12 years after repair. The reported prevalence of aortic arch obstruction (gradient higher than 30 mmHg) varies from 20% [5,85] to 27% [7]. In one series the incidence is as high as 47% [31]. Factors significantly associated with an increased risk of intervention for residual or recurrent arch obstruction include use of PTFE material, inadequate excision of ductal tissues and complex associated anatomy [5,13,15,31].

Sell et al. [31], in contrast to the findings of Tlaskal et al. [5], reported a higher incidence in patients with a direct aortic anastomosis than in patients in whom an interposition graft had been used [31]. Others reported that surgical technique for aortic arch repair is not a risk factor for development of recurrent arch obstruction [7].

Type B IAA is associated with an increased prevalence of arch reinterventions [15,32]. It was postulated that interruption of the arch proximal to the left subclavian artery increases the degree of aortic mobilisation necessary to achieve a tension-free anastomosis [15]. The higher rate of aortic arch obstruction in this morphologic subtype can therefore be related to inadequate mobilisation of the descending aorta at the time of initial repair [15].
7.4.1.1. Therapeutic options for arch restenosis. Balloon angioplasty represents the method of choice in recurrent aortic arch stenosis [1,5,7,16,31,32,85]. However, recurrence rate even after balloon angioplasty is an area of concern [1,13,15,16,50,86] and the procedure is not free of complications [1,15,16]. Some patients can require several percutaneous interventions [15].

Different surgical methods can be used as described by Monro et al., and others [5,87]. Resection of a stenotic segment with end-to-end anastomosis represents the preferred procedure which is, however, not always possible [5]. Construction of an extra-anatomic bypass and subclavian or carotid turndown procedure or patch enlargement of the stenotic area are other alternatives [1,5,7,16].

7.4.2. Bronchial compression

Usually, it develops soon after the primary repair as a result of inadequate mobilisation of the aorta and construction of a direct aortic anastomosis low and under tension [5,7,85–88]. It leads to shortening and pulling down of the arch on to the left main bronchus. The second possible cause can be the presence of residual heart lesions leading to cardiomegaly, dilatation of the left atrium, pulmonary trunk or even the aortic root [34].

It can cause prolonged ventilator dependence or refractory left lower lobe collapse [10,15,34]. The diagnosis can be made by bronchoscopy and/or by magnetic resonance imaging [34]. Reinterventions for relief of bronchial compression include aortopexy and placement of an aortic interposition graft to release aortic arch tension [7,15,85–88]. Prevention was the key and the underlying principle was the construction of a tension-free anastomosis [7,15].

7.4.3. Left ventricular outflow tract obstruction

The recurrence of significant SAS (peak gradient 25 mmHg) after repair of IAA has ranged from 17% to 67% in most series [6,11]. Sell et al. [31] reported a 42% incidence of development of this complication within the first 3 years after both the primary and the two-stage repair. However, Serraf et al. [7] reported a surprisingly low prevalence (3.6%) of LVOTO during a 10-year follow-up after primary or two-stage repair of IAA. The difference between the two studies can be partially explained by a different surgical strategy in Boston and Paris and the fact that the two series belong to different eras [4,5].

The preoperative echocardiographic parameters useful in predicting the occurrence of SAS after repair of IAA-VSD are cross-sectional subaortic area, single ventricle, type B morphology of IAA, and presence of an aberrant subclavian artery [5,59,60,89]. The predictive ability of the last two criteria for development of SAS was questioned by Luciani et al. [6]. Salem et al. [51] showed that on echocardiographic measurements an absolute diameter of the aortic valve annulus less than 4.5 mm and a Z score less than –5 predicted the development of LVOTO obstruction after arch repair, whereas size of the subaortic region did not [13,51,52]. Geva et al. [89] found an indexed cross-sectional area of the LVOT less than 0.7 mm/m² but not aortic valve dimensions to predict recurrence of LVOT obstruction.

Absence of a large VSD and initial transcatheter balloon dilation of the LVOT are associated with a significant risk of second LVOT procedure [13]. Although balloon dilatation can be an effective method of addressing isolated aortic valve stenosis, it is inadequate to address aortic valve hypoplasia and subaortic obstruction, which are more likely in the setting of IAA [13]. The persistence of the unresected conal muscle can result in sustained turbulent flow leading to progressive hypertrophy of the displaced septum and recurrent SAS [6]. It is also postulated that a complete one stage repair can reduce the incidence of late LVOTO by forcing the entire cardiac output through LVOT early in life [13,32].

These patients will require one or more additional surgical procedures to relieve the obstruction [6]. Myectomy of the conal septum from the right atrial or ventricular approach [4,5,56] and placement of a patch for VSD closure to the left side of the conal septum [6] can prevent recurrence of LVOT obstruction [1,6,16,44].

7.4.4. Pulmonary artery reintervention

The overall time-related freedom from any pulmonary artery reintervention is 78%, 75%, and 72% at 1, 5, and 10 years, respectively [13]. Reinterventions are required to relieve pulmonary artery stenosis [5,13]. It consists of balloon dilatation of the right pulmonary artery with or without stent placement and patch arterioplasty [4,5,15]. Some patients will require repeated percutaneous procedures while some patients would require bilateral pulmonary artery patch arterioplasty [15]. The use of conduits for right ventricular to pulmonary artery continuity is an important risk factor for further operations [47].

7.5. Residual heart lesions

Residual VSD requiring reoperation is described by several authors [4,5,7,90–92]. However, small VSD can close spontaneously [4,5]. Valvular aortic stenosis can develop after operation in patients with a bicuspid aortic valve and in patients who have a narrow ascending aorta and/or subaortic region [4,5]. A degree of aortic regurgitation is very common in all patients with associated TA repair [4,5,92]. Following correction of TA with IAA, right pulmonary artery stenosis can develop [5]. Primary repair of IAA combined with arterial switch operation for TGA can predispose to development of valvular and supravalvular pulmonary stenosis [5,92].

7.6. Risk factors for adverse outcome

Despite improvements substantial operative mortality persists, and a number of late complications have been reported [1,5,7,16,33]. However, more than 50% of these patients can be expected to maintain normal lives without handicap or significant limitations [4,5]. A multi-institutional prospective study undertaken by the Congenital Heart Surgeons Society identified the risk factors for death as low birth weight, younger age at repair, IAA type B, major associated cardiac anomalies, outlet and trabecular ventricular septal defects, smaller size of the ventricular septal defect, subaortic narrowing and use of a Damus–Kay–Stansel-type repair [1,8,13,16].

These conclusions are not supported by a retrospective study by Serraf et al. where none of the demographic,
anatomic, and surgical variables analysed were found to be statistically significant risk factors for early death [7]. Serraf et al. [7] reported only preoperative renal function and single-dose cardioplegia as risk factors for death. This difference can be due to the small sample size in the series by Serraf et al. [7], which emphasises the necessity of a multi-institutional study for rare anomalies such as IAA [7,16].

Tláskal et al. [4,5] reported risk factors for early mortality as poor preoperative clinical condition, acidosis, and earlier year of surgery, with better survival for those undergoing primary repair rather than palliation [13]. Female sex is associated with higher mortality in some studies [1,13].

Failure to address LVOT obstruction at the time of repair is recognised as an important predictor of mortality and reoperation [1,16,31,56]. In the presence of a small LVOT, surgeons tend to bypass the SAS by a Damus–Kaye–Stansel-type repair or to resect the SAS (myotomy or myectomy) [8,13]. Both these approaches are associated with significantly higher risk of death [8,13]. Features and management of the VSD can have a significant impact on the outcome [5,8]. Malalignment of the VSD, small size of the VSD, and the presence of multiple VSDs are risk factors for death and a marker for the presence of associated complex cardiac anomalies [5,8,13].

7.7. Early mortality and midterm results

IAA used to be a highly lethal condition with limited success in individual cases [13,93]. Neonatal operative experience, early intubation, and prostaglandin infusion have contributed to improved outcomes [5,7,33,51,52]. In the current era, in some centres the early mortality for a neonate with IAA and VSD approaches 5–10% [1,16] with a 5-year survival of 63% [5,8] to 90% [1,16]. However, association with SAS and complex malformations reduces the 5-year survival to 60% while in some other reports association of complex malformations with IAA had no statistical impact on early and late mortality [7]. Reported survival at 10 years varies from 47% [13] up to 85% [47,91].

The clinical focus has now moved from early operative survival to medium- and long-term outcomes [7,13,33,51]. The long-term outcome is dominated by the need for reoperation and/or reintervention despite significant advances in recent years [1,16]. The clinical development may also be affected by the presence of co-existent non-cardiac diseases such as respiratory and gastrointestinal problems [4,5,7,8,85,91]. Neurological and psychomotor developments also get adversely affected [85,91]. Signs of chorea and other CNS functional lesions may occur relatively late after surgery with a hypothermic low flow bypass or circulatory arrest [4,5,91]. The long-term influence on neurological and psychomotor development will require further studies.

7.8. Conclusion

Management of IAA and associated anomalies remains a surgical challenge. Recent improved results are a result of better understanding of the anatomy, better preparation of candidates for operation, and more intensive preoperative resuscitation of the patients. Single-stage repair provides satisfactory medium-term results though the debate continues over its advantages over staged approach particularly in association with complex anomalies. Long-term results are dominated by repeated reinterventions for LVOT and arch obstructions. Large multicentre studies are needed to resolve several unanswered questions.

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