

Anaesthesia recommendations for **Bland-White-Garland syndrome**

Disease name: Bland-White-Garland syndrome

ICD 10: Q24.5

Synonyms: BWGS, Anomalous left coronary artery from the pulmonary artery (ALCAPA), White-Garland syndrome

Disease summary: The Bland-White-Garland syndrome (BWGS) is an extremely rare congenital cardiac abnormality [28]. This syndrome was first mentioned in 1886, but the full anatomy and clinical symptoms were first described by Bland, White and Garland in 1933 [5, 6].

Within this syndrome, the left coronary artery (LCA) has an unexpected anomalous origin from the pulmonary artery instead of the aorta. Blood flow in the coronary system follows the direction of pressure gradient and takes the path of lowest resistance [14,15]. In patients with BWGS, blood flows from the normal origin of the (often enlarged and dilatated) right coronary artery (RCA) through myocardial coronary collateral vessels to the LCA and low-pressure system of the pulmonary artery system [15,25]. After the drop of pulmonary vascular resistance in the first months of life, the majority of the blood drains to the pulmonary arteries, causing a “coronary steal” phenomenon, a left-to-right shunt and ultimately leads to an abnormal left ventricular perfusion [35]. This may lead to myocardial ischaemia and infarction, left ventricular dysfunction, mitral insufficiency and regurgitation, congestive heart failure and finally to sudden cardiac death [35,47].

BWGS occurs between 0.26 % and 0.46 % of patients with congenital heart disease and appears once in 300,000 births [3,22]. It is usually seen as an isolated cardiac abnormality, but in about 5 % of cases, it appears in coexistence with other cardiac abnormalities like patent ductus arteriosus (PDA), atrial septal defect (ASD), ventricular septal defect (VSD) with or without mitral stenosis, tetralogy of Fallot, pulmonary stenosis, coarctation of the aorta or transposition of the great vessels [1,4,8,10,15,34,35,43,45,48].

Symptoms usually become clinically apparent shortly after the neonatal period [15]. Vascular resistance is equal in right (pulmonary artery) and left (aorta) system during foetal period. This condition allows the myocardium supplied by the anomalous artery to remain well perfused. Promotive at this stage of development, there is an identical oxygen content in the pulmonary artery in comparison to the aorta. For this reason, this coronary anomaly is well tolerated in foetal and early neonatal life [4,35,38]. These conditions lead to an antegrade flow in anomalous LCA as well as in normal RCA [35]. In consequence, coronary collateral growth is not especially promoted before birth [38]. When pulmonary arterial pressure decreases physiologically after birth, the antegrade LCA flow diminishes and reverses. Moreover, the antegrade LCA is filled with deoxygenated mixed venous blood [4]. Subsequent, this coronary steal phenomenon leads to ventricular (anterolateral) myocardial ischaemia and in further consequence to mitral valve papillary muscle dysfunction [15,35].

Clinical presentation of BWGS may vary but primarily goes along with symptoms of heart failure like angina-like symptoms, severe dyspnoea, tachypnoea, cyanosis, wheezing, pallor, dizziness, profuse sweating, signs of decreased peripheral perfusion, syncope and pulmonary oedema or malignant arrhythmias [13,15,17,40,47]. In children, a failure to thrive, a smaller stature, diaphoresis while drinking and feeding, reduced exercise tolerance or poor weight gain may occur [15,51].

Two types of BWGS can be differed [25,35]. The infant type applies to patients without collateral vessels. Regarding differential diagnosis in this age, dilatated cardiomyopathy of differing genesis should be considered [35].

In contrast, the adult type occurs in patients with well-established collateral vessels and possible coronary ostial stenosis. These collaterals between RCA and LCA were formed during childhood due to ischaemic stimulation. They are essential and the only way to survive beyond infancy without surgery. Despite formation of these collaterals, adult patients develop a chronic left ventricular subendocardial ischaemia over the years, with high risk of lethal ventricular arrhythmias [35]. Asymptomatic patients with BWGS presenting in adulthood are rare [21,24,28,29,44,50,51]. They must have a well-developed coronary collateral circulation with retrograde perfusion of the left ventricle from the RCA [28,38]. An ostial stenosis of the LCA or persistence of elevated pulmonary resistance might limit the steal and increasing myocardial perfusion pressure. At least the degree of collaterals determines the moment of medical consultation due to beginning cardiac decompensation [15]. Nevertheless, adult BWGS patients without performed surgical correction are at high risk for sudden death. Typical is a precipitation by exercise at an average age of 35 years in untreated patients with BWGS [25,38,50].

The definitive treatment option for BWGS is cardiac surgery [49,51]. Untreated, 90 % of patients with BWGS die within the first year of life [35,47]. In case of early diagnosis and prompt surgical intervention, patients with BWGS may achieve adulthood. This applies also for asymptomatic patients [25]. There is one case report of an 88-year-old woman with BWGS, but, due to severe cardiac impairment, this expectation of life remains a rarity in this disease [42]. Supportive and conservative measures consist of typical pharmacotherapy of congestive heart failure (e.g., diuretics, β -blockers and in acute failure: inotropes) [12,18,44].

Medicine is in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong



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Emergency information

A	AIRWAY / ANAESTHETIC TECHNIQUE	no special airway malformations – GA (TIVA / balanced) may induce (rapid / severe) myocardial decompensation – therefore, consider peripheral RA as a feasible alternative if applicable – consider haemodynamic deterioration after neuraxial blockades (reduction of cardiac afterload) – avoid hypocarbia, hyperoxia and alkalosis to maintain high PVR
B	BLOOD PRODUCTS (COAGULATION)	no specific recommendations
C	CIRCULATION	anamnesis: coronary correction surgery performed? – anticipate coronary steal phenomenon = left-to-right shunt → abnormal LV perfusion → (chronical) myocardial ischaemia with high risk of lethal ventricular arrhythmias, LV dysfunction, mitral insufficiency / regurgitation, congestive heart failure and SCD – coexistence with other anomalies feasible, i.e. PDA, ASD, VSD with(out) mitral stenosis, tetralogy of Fallot, pulmonary stenosis, coarctation of the aorta or transposition of the great vessels – supportive measures consist of typical pharmacotherapy of congestive heart failure (e.g., diuretics, β-blockers and inotropes) up to LVAD or ECMO – consider IBP and (non)invasive haemodynamic monitoring (preload? CO? SVR?) or FoCUS
D	DRUGS	no risk for MH – avoid epinephrine as adjunct in local anaesthetics (potentially haemodynamic deterioration) – endocarditis prophylaxis (if indicated)
E	EQUIPMENT	anticipate myocardial decompensation within extreme positioning

Typical surgery

Being the only curative therapy, surgical correction is considered to be standard and definitive treatment [15,35]. In nearly all children, cardiac surgery is indicated and should be performed before the development of severe myocardial dysfunction [15]. The aim of surgery is to restore a two-coronary-artery circulation system [13,35]. Especially in patients with infant type of BWGS, early cardiac surgery is essential to prevent myocardial ischaemia, malignant arrhythmias, sudden cardiac death and to keep ventricle function adequate [28,35].

There are two main approaches in correction surgery.

Aspiring a one-coronary system includes ligation of the anomalous LCA at its pulmonary origin. With this approach, left ventricular coronary blood flow is only based on RCA collateral circulation whereas the low-oxygenised blood flow from the pulmonary artery is stopped. This procedure contains a greater risk for complications and is therefore usually avoided [15,28,35].

Restoring a two-coronary system is currently advocated and includes the direct re-implantation of the LCA into the aorta, the creation of a transpulmonary baffle between the coronary ostium in the pulmonary artery and the aorta (Takeuchi procedure), the placement of a coronary artery bypass graft (CABG) combined with ligation of the origin of LCA or saphenous vein LCA bypass graft into the proximal aorta preserving a dual coronary arterial system [15,28,35].

The latter method is preferred in adults, the former one in children [35].

Depending on the degree of improvement in impaired mitral valve function after correction surgery, a reconstruction or replacement of mitral valve may be necessary [26].

In patients with progressive left ventricular dysfunction, temporary therapeutic regimes may include the implantation of left ventricular assist device (LVAD) and extracorporeal membrane oxygenation (ECMO). Both procedures may restore myocardial function, reduce mortality, and serve as a bridge to heart transplantation or recovery [2,23,30,43].

As last resort and in cases of severe heart failure, cardiac transplantation might be necessary.

Type of anaesthesia

The anaesthetic technique influences myocardial perfusion and should therefore be handled with great care. With progression of myocardial dysfunction, oxygen consumption should be reduced as much as possible during induction and maintenance of anaesthesia. A decrease in preload caused by most anaesthetics due to vasodilatory effects as well as a decreased systemic vascular resistance and increased venous capacitance (that will limit ventricular filling) may severely limit cardiac output and at least exacerbated ischaemia may further compromise the left ventricular function [37]. Each negative inotrope substance should be titrated slowly and carefully to avoid increased catecholamine response [15].

A sufficient RCA perfusion due to an adequate diastolic blood pressure should be aspired to ensure myocardial perfusion in patients with BWGS. An unnecessary high systemic vascular resistance / afterload might increase left ventricular end-diastolic pressure and finally result in diminished coronary perfusion. This tightrope walk, a dysfunctional and impaired ventricle function with low afterload and simultaneously adequate diastolic blood pressure, is the main challenge in patients with uncorrected BWGS [15].

For premedication midazolam or diazepam are recommended by some authors to improve haemodynamic stability before induction [9,23].

There are only few case reports on anaesthesia in patients with BWGS. There is one case report on a patient with necessity of CPR after induction with sevoflurane, fentanyl and pancuronium. Other case reports refer to the use of these substances as uneventful as well as usage of midazolam, ketamine, etomidate, propofol, remifentanyl, morphine, rocuronium and vecuronium [4,9,15,18,20,23,30]. Another case report on a patient with undiagnosed BWGS undergoing emergency laparotomy refers electromechanical dissociation and necessity of CPR after induction with propofol, fentanyl, succinylcholine and cisatracurium [12]. Years later, circulatory arrest occurred again in the same patient during the induction of general

anaesthesia (with midazolam, fentanyl, propofol and rocuronium) due to ventricular tachycardia and asystole [12].

Despite known side effects like adrenal suppression, some authors recommend etomidate instead of propofol as hypnotic substance for induction [12]. However, the careful titration of selected hypnotic drugs is more important than the dedicated use of one specific agent without adequate titration in this specific patient population.

In case of unavailability of an intravenous catheter, sevoflurane gas induction was performed [15]. With respect to reports of possible cardiopulmonary resuscitation after use in patients with dilatative cardiomyopathy, intramuscular application of ketamine was proposed as another alternative for induction in patients with BWGS [15, 37]. Intranasal application may also be an alternative, especially in children.

For maintenance of anaesthesia, continuous infusion of midazolam and fentanyl were reported as uncritical as well as sevoflurane and isoflurane, whereas the latter one is preferred by some authors [15,18,20,23].

Pulmonary vascular resistance is most likely raised in patients with BWGS. Caution should be exercised during ventilation to avoid lowering pulmonary vascular resistance further with high inspired oxygen, hypocapnia and alkalosis. This may lead to a progress in coronary artery "steal" [15].

Impaired ventricle function may require inotropic support to stabilise patient's haemodynamics [23]. Subject to the particular situation, the use of adrenaline, noradrenaline, dopamine, dobutamine, epinephrine, phenylephrine, milrinone and enoximone were used to stabilise patients [15,23]. Nevertheless, inotropes should be used cautiously, as they increase the consumption of myocardial oxygen, which may accentuate myocardial ischaemia [18].

In case of ventricular fibrillation, lidocaine was reported as effective besides defibrillation [15]. Nevertheless, the common guidelines for cardiopulmonary resuscitation with necessary defibrillation should be followed.

Haemodynamic consequences of decreased myocardial perfusion can occur even using a careful inhalation anaesthetic technique in these critically ill patients. There is no strong recommendation whether intravenous or volatile induction and maintenance should be preferred [15,20]. This decision should include the local experience and preference of the team members.

For additional analgesia, paracetamol was reported as uneventful [20].

Regional anaesthesia may be an alternative for undergoing suitable operations. Avoiding the effects of general anaesthesia on haemodynamic, peripheral regional anaesthesia should be preferred for extremity surgery. Neuraxial blockades might be feasible, however, the potential severe reduction of cardiac afterload due to vasodilatation in neuraxial blockades (spinal / epidural / paravertebral anaesthesia) might result in challenging acute haemodynamic deterioration. The use of ropivacaine for paravertebral block was reported as uneventful in a patient with BWGS [20]. There is no recommendation for a specific type of local anaesthetic drug. Epinephrine as an adjunct in some local anaesthetic formulation should be avoided due to the risk of systemic resorption and unwanted and potentially deleterious effects on the myocardium.

Necessary additional pre-operative testing (beside standard care)

There is no general recommendation or protocol for an ideal preoperative assessment. Due to congestive heart failure, other organs may be dysfunctional. Therefore, a consequent pre-operative assessment in which specific symptoms and organic dysfunction must be identified in the individual patient is indispensable. Especially an impaired function of heart, lung, kidney, and other organs should be optimised and controlled as far as possible in pre-operative management.

A thorough clinical evaluation and anamnesis may help to assess the patient's current functional status and to plan further pre-operative diagnostic.

A 12-lead ECG should be induced pre-operatively. Due to an impaired perfusion of left coronary artery, sometimes alterations compatible with infarction of the anterolateral wall are observed in patients with BWGS [32,41]. Precordial leads V1-6 as well as leads I and aVF may show pathological ST-segment variations [41,52]. Furthermore, Q-waves, left ventricular hypertrophy and left axis deviation may be seen [50]. Atrial fibrillation was also associated with BWGS [17,31].

In case of impossibility of a treadmill stress test for deliberate provocation, a myocardial scintigraphy may demonstrate areas of ischaemia [12].

A chest X-ray may identify cardiomegaly, one of the most common presenting features in BWGS [15,23].

Transthoracic (TTE) or transoesophageal (TEE) echocardiography may help identifying BWGS non-invasively and evaluating the function of cardiac valves [15,46]. Criteria for diagnosis of BWGS include identification of a dilated RCA, retrograde Doppler flow from the LCA to pulmonary artery and prominent septal flow from collateralisation [16,49]. Besides, mitral valve regurgitation, dilatation of the left ventricle as well as hypokinesia of the anterolateral wall may typically be observed [16].

Diagnosis can be confirmed or excluded by left cardiac catheterisation or coronary angiography as gold standard investigation for diagnosis. Typical is a markedly dilated, aneurysmal, and tortuous RCA [36].

Magnetic resonance angiography (MRA) and multislice computed tomography (CT) provide non-invasive visualisation of the coronary anatomy and may be performed as well. Both techniques have surpassed the X-ray coronary angiography and are recommended for diagnosis and characterisation of coronary anomalies [12,15,18,23,35,44].

Especially infants presenting for (non)cardiac surgery showing evidence of unexplained myocardial ischaemia on ECG peri-operatively, associated with left ventricular dysfunction, should have the diagnosis of anomalous coronary artery origin, and be treated with a high index of suspicion [15].

Of course, report on any of the typical warning symptoms for congestive heart failure in adults should raise suspicion of heart disease, and specific tests should be considered even in low-risk patients, especially when a major surgery is being planned [12].

Even in asymptomatic patients, caution should be exercised because of few case reports referring pre-operative evaluation and laboratory tests as inconspicuous (except coronary angiography). However, during induction of general anaesthesia, the patient became haemodynamically instable and underwent CPR subsequently [12].

Particular preparation for airway management

Airway management, especially laryngoscopy and intubation, should be performed smooth and rapid to avoid stress and increased catecholamine response with risk of myocardial decompensation [18].

Massively enlarged heart silhouette may compromise respiratory function and aggravate ventilation. There is one case report in an infant with BWGS, in which the left main bronchus was compressed by the enlarged left atrium. Consequently, atelectasis of the left lung led to respiratory failure. Stenting of the left bronchus was necessary to ensure effective ventilation [39].

As usual in planning anaesthesia, alternative strategies for airway management should be planned in advance if difficulties in airway management are presumed.

Particular preparation for transfusion or administration of blood products

No specific recommendations are given. No typical bleeding disorders were reported for patients with BWGS. For cardiopulmonary bypass, heparin was commonly used [15].

Particular preparation for anticoagulation

There are no specific suggestions for BWGS. Subject to cardiac and valvular surgery, arrhythmias, strokes or other cardiovascular events in the patient's anamnesis, anticoagulation should be considered after operation corresponding to current recommendation.

Particular precautions for positioning, transportation and mobilisation

Extreme positioning for specific operations, e.g., a (reverse) Trendelenburg positioning or prone position, might lead to haemodynamic impairment in case of severe cardiac involvement. Depending on remain of myocardial function, patients may decompensate rapidly if patient's positioning influences increased changes in cardiac pre- or afterload.

Interactions of chronic disease and anaesthesia medications

Not reported. Supportive pharmacotherapy for management of congestive heart failure should be continued corresponding to general recommendations for pre-operative assessment in general anaesthesia.

Anaesthetic procedure

Because of limited experience of anaesthesiologists in the management of this rare cardiac malformation, it is difficult to make specific recommendations [23].

Patients with uncorrected coronary status are commonly treated by paediatric cardiac surgery teams and paediatric cardiac anaesthetists who are experienced with BWGS syndrome and its perioperative challenges. In all other uncorrected cases with scheduled non-cardiac procedures, consultation with paediatric cardiologists (or cardiac specialists for adults with congenital heart disease for adult patients) is strongly recommended.

Pre-operative evaluation: see details above.

Premedication: might be performed weighing the benefits and risks in individual patients.

Prophylaxis for endocarditis: should be performed on patients with an indication for prophylaxis (mainly after cardiac valve surgery) according to current international guidelines and after discussion with the responsible cardiologist [19].

Patient positioning and monitoring: act with caution due to haemodynamic impairment in case of severe cardiac dysfunction and haemodynamic instability.

Vessel cannulation: multiple attempts of intravenous cannulation as well as other stressing factors should be avoided to prevent catecholamine response and possible resultant myocardial ischaemia [15].

Non-invasive monitoring: should include 12-lead ECG with real-time ST-segment analysis, pulse oximetry and blood pressure [15,20]. Ventilatory settings, especially oxygen saturation and end-tidal carbon dioxide should be monitored very closely [20]. To maintain a high pulmonary vascular resistance in patients with BWGS, hypocarbia, hyperoxia and alkalosis should be avoided [18].

Anaesthesia: induction of anaesthesia should be performed under consideration of patient-specific risk factors and to turn attention to cardiac impairment. Total intravenous or balanced anaesthesia using volatile anaesthetics can be performed with established drugs (see details above). Due to usually severe impairment of myocardial function and few case reports of sudden cardiac arrest during anaesthesia in patients with BWGS, the anaesthesia team should continuously be prepared for CPR during the whole procedure of anaesthesia and surgery [12, 15].

Severity of cardiac impairment may necessitate the use of regional anaesthesia techniques in patients with BWGS. Nevertheless, there are rarely case reports referring this condition [7].

Particular or additional monitoring

Usually, anaesthesiologic monitoring should be performed according to the principles for patients with impaired myocardial function and patients undergoing cardiopulmonary bypass [23].

Invasive monitoring may comprise arterial line for blood pressure, central venous catheter with measurement of central venous pressure [15].

If available, cardiac output (CO) monitoring may help to regulate fluid management and catecholamine therapy. Ensuring an optimal preload and myocardial contractility as well as a low or normal systemic vascular resistance (respectively a low afterload) and to prevent a decrease in pulmonary vascular resistance or tachycardia are important anaesthetic goals in taking care of patients with BWGS [18]. Probably a regular heartbeat within normal ranges may be ideal. Despite a prolonged diastole, bradycardia may lead to a paradoxical blood flow

due to a steal phenomenon. Theoretically, this may increase oxygen requirement of the left ventricle and lead to ischaemic impairment and a decreasing oxygen supply in this coronary anomaly [33]. In these patients, monitoring the stroke volume is advantageous to measuring the CO, because CO varies with heart rate [18].

An additional non-invasive method to monitor cardiac function is focused cardiovascular ultrasound (FoCUS). This is suggested as routine use in cardiac impaired patients and can be performed pre-operative, during induction as well as at the end of surgery on intensive care unit or in the holding area to reconfirm the absence of disturbances in myocardial function [11,20]. Intraoperative TEE is used in cardiac procedures according to international guidelines whenever applicable [15].

Possible complications

Depending on the severity of patient's myocardial impairment and due to potentially rapid myocardial decompensation, cardiac support by techniques like extracorporeal membrane oxygenation (ECMO) or left ventricular assist devices (LVAD) may be necessary [15,30].

In case of additional cardiac anomalies like ASD or VSD, the changing pressure ratios after their operative closure may be challenging. If no pre-operative diagnosis of BWGS was available, the repair of these anomalies may unmask undiagnosed asymptomatic BWGS. VSD closure for example caused in one case a concomitant decrease in pulmonary blood flow and led to the development of LCA "steal phenomenon" that had not been present while the VSD was open [15].

After ligation of a PDA, ventricular fibrillation occurred in a patient, unmasking an undiagnosed BWGS [4].

Sudden cardiac arrest in general anaesthesia may be the first manifestation of BWGS in affected patients as well as in undiagnosed patients [12].

Post-operative care

Post-operative care should be based upon the intervention and the patient's pre-existing (myocardial) conditions.

Because the main reason for undergoing surgery in patients with diagnosed BWGS is an operative correction of the coronary anomaly, patients are treated initially on ICUs post-operatively.

Especially patients with corrected coronary anomaly of BWGS who were haemodynamically stable during non-cardiac interventions may be monitored for a prolonged time interval in the anaesthetic recovery room. Transfer to intermediate or intensive care units is not mandatory but might be reasonable if severe myocardial dysfunctions exist or if there is any sign of haemodynamic instability.

An adequate pain control with a multimodal analgesia regime in the intraoperative and immediate post-operative period is essential for patient's comfort as well as a rigorous maintenance of core temperature during the procedure to obviate gratuitous stress [20].

Disease-related acute problems and effect on anaesthesia and recovery

There are few differential diagnoses for dilatation of the coronary arteries. This includes e.g., non-ischaemic cardiomyopathy of different genesis, critical coarctation of the aorta, severe aortic stenosis, Kawasaki disease, coronary artery-coronary sinus fistula, atherosclerosis-related coronary artery ectasia, vasculitis (polyarteritis nodosa or Takayasu arteritis), scleroderma, Ehlers-Danlos syndrome, hereditary haemorrhagic telangiectasia, myocarditis, trauma and hyperlipidaemia [15,35].

Haemodynamic deviation may also occur due to pulmonary embolism, anaphylaxis, bleeding complications up to haemorrhagic decompensation or desaturation and hypoxia because of airway affection.

Ambulatory anaesthesia

Ambulatory anaesthesia is possible in patients with corrected BWGS and may be performed in institutions with adequate resources and expertise. This strongly depends on the type of surgery, intra-operative haemodynamic stability and on the patient's post-operative myocardial function. This may be evaluated by bedside TTE, ideally with respect to the patient's pre-operative status [20].

Uncorrected BWGS patients should not undergo ambulatory anaesthesia.

However, there are no general recommendations regarding outpatient procedures due to a lack of existing literature.

Obstetrical anaesthesia

Because the majority of patients with BWGS does not achieve adulthood, there are rarely case reports of pregnant women with diagnosed BWGS. In general, women with BWGS are fertile. Thus, obstetrical anaesthetist might face pregnant women for vaginal deliveries or Caesarean section [21,25,50,51]. In one case report, termination of pregnancy was recommended for undergoing repair surgery due to severe cardiac impairment [52]. Two other case reports refer successful pregnancy and vaginal delivery in women who underwent repair surgery in childhood. Both were closely monitored during pregnancy with clinical inspection and TTE in constant ranges until birth [7]. One of these two women had epidural analgesia for childbirth, and no complications were reported.

Supportive pharmacotherapy with diuretics or antihypertensive substances after conception is often terminated due to risk for foetal development. This may lead to progress in myocardial dysfunction in pregnant women.

One case report refers a successful emergency Caesarean section due to sudden cardiac decompensation in a pregnant woman with preeclampsia and undiagnosed BWGS [27]. Preeclampsia can lead to extensive systemic vasoconstriction, haemo-concentration and increased blood viscosity which may lead to myocardial ischaemia even in women with a susceptible cardiac lesion [27].

However, given the rarity of this condition (pregnancy in patients with BWGS), specific guidance and recommendations regarding management cannot be given [7].

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