Functional evaluation of the upper extremities after corrective surgery of congenital defects of the aortic arch in children depending on arterial vascularisation

Ocena funkcjonalna kończyn górnych po operacjach naprawczych wad wrodzonych łuku aorty u dzieci w zależności od unaczynienia tętniczego

Agata Wasilewska¹, Jakub Wasilewski¹, Paulina Ewertowska¹, Natalia Szulc¹, Konrad Paczkowski², Katarzyna Gierat-Haponiuk¹, Julia Haponiuk-Skwarlińska², Afrodyta Zielińska¹, Jolanta Zajt¹, Ireneusz Haponiuk¹, ²

¹Department of Health and Biological Sciences, Jędrzej Śniadecki Academy of Physical Education and Sport in Gdańsk, Poland
²Department of Pediatric Cardiac Surgery, St. Adalbert in Gdańsk-Zaspa, COPERNICUS PL Sp. z o.o., Poland

Abstract

Congenital heart defects are often accompanied by defects in great vessels, such as aortic coarctation with various forms of aortic arc hypoplasia, vascular rings and positional anomalies. Surgical treatment and the choice of optimal defect correction technique in the child may affect the restoration, maintenance or the arrest of blood supply by the aorta to arterial vessels given off by the aortic arch. Proper, closest physiological vascularisation of individual parts of the body and organs is necessary for proper development and maintenance of physiological function. The consequence of the use of various strategies for correcting congenital defects of the aorta and the use of available surgical techniques may be perfusion disorders, also in the field of vessels supplying upper limbs, most often the upper left limb.

In this article, we have attempted to develop a universal strategy for assessing the development and function of the upper extremities in children. It consists of cardiac methods of cardiovascular evaluation and physiotherapeutic function tests of the upper limbs adapted to the age of patients. Confirmation of the long-term effects of maintaining or restoring the blood supply by the aorta on the basis of objective function tests of the upper extremities may be an additional argument while choosing the optimal surgical method in the youngest children.

Key words: congenital heart defects, defects of the vessels of the aortic arch, pediatric cardiac surgery, coarctation of the aorta, hypoplasia of the aortic arch, cardiosurgical correction, evaluation of the upper extremities, muscle strength, EMG, AHA

Introduction

In this article we attempt to develop a universal strategy for assessing the development and function of the upper extremities in children. It consists of cardiac methods of cardiovascular evaluation and physiotherapeutic function tests of the upper limbs, adapted to the age of patients. Confirmation of the long-term effects of maintaining or restoring the arterial blood supply originating from the aorta on the basis of objective function tests of the upper extremities may be an additional argument for choosing the optimal surgical method in the youngest children.
Congenital heart and vascular abnormalities also concern structural abnormalities of vessels giving off by the aortic arch. This can cause abnormal blood flow and give clinical symptoms in the area of their atypical pattern and vascularisation, including the organs of the mediastinum, upper extremities and the head. The aim of surgical treatment of congenital heart defects is to correct all reparable anomalies that can be corrected. As a result, blood circulation is as close to the physiological standard as possible, regardless of the extent of primary, congenital defect. The correction of congenital defects also includes corrective procedures of the main artery and vessels giving off by its arch, i.e. vessels supplying the head. Vascular intervention usually changes their spatial orientation and position as well as in the blood supply of the corrected area. This fact may be important for the further functioning of both the arteries as well as the structures and organs vascularised by them.

The most common vascular-related congenital anomalies of the aortic arch include coarctation of the aorta with various forms of aortic arch hypoplasia, vascular rings and positional anomalies of the aorta and pulmonary artery in the mediastinum. The surgical technique selected for the treatment of these defects may affect the maintenance or restoration of the blood supply from the aorta to the originating arteries, which is possible by changing its position, functional purpose, and spatial orientation. This may require an additional corrective procedure.

Traditional surgical strategies for the correction of congenital anomalies of the heart and mediastinal vessels have been developed to improve arterial blood circulation in the basic vascular structures, which is the main goal. The complex methods of the blood supply reconstruction to all originating vessels require modification of the surgical technique with an extension of the surgical field, duration of the procedure and degree of the intervention into the thoracic cavity. The patient’s early age, i.e. neonatal period and infancy makes it even more difficult to make the decision to change the surgical strategy. Technical aspects such as low body weight and small size of a child may also cause difficulties. This is in contradiction to the rational premise that the simultaneous correction of all the reparable defects may have a positive impact on the child’s further development.

Assumptions

Proper arterial vascularisation of tissue and organ is based on proper development and maintenance of physiological function. The maintenance of physiological vascularisation of the shoulder girdle and upper extremities in patients after cardiovascular correction procedures should therefore guarantee the proper development and function of these regions.

The anatomy of the defect and the way it is corrected cause hemodynamic effects, both on the total blood circulation in the body and on the blood flow in the carotid and vertebral vessels, directly adjacent to the region of the coarctation of the aorta and the hypoplastic aortic arch. The consequences of the use of various strategies for correcting the aortic coarctation and the use of a specific surgical technique, frequently with necessary arrest of blood flow, may include perfusion disorders, also in the field of vessels supplying upper limbs, usually the left upper limb.

As a result of the aortic arch anomaly with a limited blood supply originating from the aorta to the left subclavian artery, which is operated on in children (frequently in a newborn), the long-term consequence of the procedure may be limitation of the functional efficiency of the left upper extremity.

Aim of the study

To assess the late consequences of maintaining vascularisation by the aorta we have attempted to develop a universal strategy for assessing the development and function of the upper limbs in children after surgery of congenital heart and vascular defects, using the methods for cardiological assessment of the cardiovascular system and physiotherapeutic tests for functional evaluation of the upper extremities, which are age-appropriate and adapted to development of examined patients.

Confirmation of the long-term importance of maintaining or restoring the arterial blood supply originating from the aorta on the basis of objective, minimally invasive function tests of the upper limbs may be an additional, important argument in the discussion about the proper choice of optimal surgical strategy in the youngest children.

The most common congenital defects of the aortic arch and great arteries given off by the aortic arch— including initial anatomy and pathophysiology

Coarctation of the aorta with aortic arch hypoplasia

Coarctation of the aorta (CoA) is a congenital heart defect defined as aortic narrowing in the area of the aortic isthmus [1]. This defect occurs in approx. 0.4% of newborns and 6–8% of children with congenital heart defects [2]. It may occur as an isolated anomaly, or may be a component of other defects, resulting in a variety of clinical symptoms and threats to the whole body. In a newborn with critical CoA, only an early start of intensive treatment ensures their survival. This defect is classified as ductus-dependent critical one.
The basic literature describes two types of anatomy of both the aorta and aortic arch, treated as physiological forms. Type I, the most common variant (74.0–89.4% of the population), consists of three great vessels emerging from the aortic arch. The first vessel is the brachiocephalic trunk, which is divided into the right subclavian artery and the right common carotid artery. The second vessel is the left common carotid artery, and the last structure branching off from the aortic arch is the left subclavian artery. The second variant is so-called ‘bovine trunk’ (a profile of a bull) and occurs in 7.2–21.1% of the population [3]. Some authors also distinguish two subtypes of the second variant: ‘long bovine trunk’, which is type II-A, and ‘short bovine trunk’, which is type II-B [4]. It should be emphasized that both variants are physiological forms, with maintaining the proper blood flow, and all elements of the proper anatomy of the vessels.

In the congenital defect of the heart and vessels, called ‘bovine trunk’, the brachiocephalic trunk and the left carotid artery give off as a common vessel in the continuation of the ascending aorta. The proximal aortic arch does not develop in this anomaly. The continuation is the distal aortic arch with a relatively long shape reaching the region of the left subclavian artery and the aortic isthmus. In the form of a neonatal defect the peripheral flow of blood to the organs situated below the aortic arch is provided by an unobstructed arterial duct.

Vascular rings with the anomaly of the pattern of subclavian arteries

The subclavian artery ensures the proper arterial vascularisation of the upper extremity, which, after branching off from the brachiocephalic trunk on the right or distal aortic arch (on the left), becomes a strong axillary artery responsible for supplying the upper limb muscles with the shoulder girdle [5]. The axillary artery starts at the outer edge of the first rib. It ends at the height of the edge of the lower pectoralis major. The axillary artery is supported by a small number of branches connecting to the carotid branches of the subclavian artery and intercostal arteries.

A heart defect called a vascular ring contains many anatomical forms. From the point of view of the upper limb vascularisation, the anomalies of the origin and pattern of subclavian arteries, which incorrectly give off by the aortic arch on the opposite side of the extremity supplied, are of great importance. Such a position of arterial vessels causes compression symptoms of the mediastinal organs, including vitally important ones: trachea, bronchi, and esophagus. In the international nomenclature, the Latin term *arteria lusoria* has been adopted, which means the (right or left) aberrant artery.

Impression (compression) of vascular structures on mediastinal organs

The position of the aortic arch with its version, referred to as either left or right one, or in an intermediate position, specified clinically as ‘zero’ one may cause compression symptoms, additionally exacerbated by the basic hemodynamic parameters of the aorta: high blood pressure, high pulse wave with high blood flow volume and stiffness of its wall.

Patients with disadvantageous position and spatial orientation of the aorta in its initial, mediastinal segment show symptoms of impression, similar to those typical of the presence of a vascular ring.

Functional muscles of the upper extremities, responsible for physiological function of the limbs

The key muscles in the upper extremity function include rotator cuff, deltoid muscle, biceps and triceps, flexor and extensor groups of the wrist and hand [6]. The above-mentioned muscles also need vessels supplied from the aorta with an appropriate diameter and capacity to function properly. Adequate arterial vascularisation also seems to be a natural condition for the physiological development of the extremities, which is consistent with the growth of the whole body during childhood.

Cardiosurgical correction techniques in the treatment of vascular defects of the aortic arch

Standard methods of repair of aortic coarctation with hypoplasia of the aortic arch

The preferred treatment strategy today is an early repair of the aorta within the limits of its own healthy tissues. Coarctation of the aorta is often accompanied by various forms of underdevelopment (hypoplasia) of the aortic arch [7]. The traditional surgical treatment consists in the resection of the narrowing region with the removal of abnormal tissues; in some techniques — with the resection of the carotid and vertebral vessels, mainly the left subclavian artery. Performing the most popular corrective surgery in newborns and infants with low body weight makes it difficult or impossible to maintain the left subclavian artery, in which the blood flow is ensured by the presence of the collateral circulation. The sources of the collateral circulation for the severed subclavian artery include the internal thoracic artery and the vertebral artery — via the Circle of Willis, and vessels in the region of the shoulder girdle. Deprivation of vascularization by the aorta for such an important vessel as the subclavian artery, which supplies large muscle parts
of the upper extremity, predisposes to the formation and the symptoms of the so-called steal syndrome due to the backflow of blood in arteries. The symptoms of the steal syndrome, which are the most difficult to compensate, include neurological complications resulting from CNS hypoperfusion in the circle of Willis. The most common early symptoms in patients include headaches, dizziness and synapses that are dangerous to life and health.

A modified technique of coarctation resection with narrowing of the aortic isthmus and proximal anastomosis by end-to-side method, with reconstruction of the blood supply to the left subclavian artery using a hypoplastic, distal segment of the aortic arch

Correction of the ‘bovine trunk’ anomaly in a newborn is possible by means of a standard lateral access. In the initial stage of the operation the blood supply to the left subclavian artery is reconstructed using a distal segment of the hypoplastic aortic arch, which smoothly changes into the left subclavian artery. By maintaining an infusion of prostaglandin and arterial duct patency, the brain and both upper limbs are supplied from heart while the lower part of the body is perfused from the duct. After ligation of the arterial duct, the descending aorta is closed with a vascular clamp and the narrowed aortic isthmus is extensively resected along with the periductal tissue, followed by — after lateral incision of the proximal part of the aortic arch at the base of the common brachiocephalic trunk and large malalignment of the descending aorta — performing the end-to-side anastomosis. An additional complication may be the anatomical variant including the subclavian artery origin in the area of the narrowed aortic isthmus or even below — from the periductal tissue, which requires independent transplantation of the left subclavian artery to the trunk of the left common carotid artery, with end-to-side anastomosis.

After this procedure with end-to-side anastomosis, the blood flow to the descending aorta is bypassed by a segment of the aortic arch and its hypoplastic structure is used only to maintain continuity and to provide blood supply originating from the aorta to the left subclavian artery. The main advantage of this technique is that it can be performed without cardiopulmonary bypass, using lateral access, with the use of healthy, patient’s own tissues and within their borders, while maintaining blood supply by the aorta to all arteries.

Clinical evaluation of a patient after aortic arch surgery

Diagnosis and imaging methods

The most important diagnostic issue is the initial anatomy of vascular anomalies. In patients with coarctation the intensity of aortic narrowing in the area of the aortic isthmus, aortic arch hypoplasia, as well as development and orientation of vessels giving off by the aortic arch are evaluated. Children with the vascular ring diagnosis require aortic arch identification and detailed evaluation of both the proximal segment of origin of carotid and vertebral vessels and their further pattern. In patients with symptoms of great vessel impression, the position of the aortic arch with its version, referred to as either left or right one, or in an intermediate position, specified clinically as ‘zero’ one, requires detailed imaging. In addition, the condition of the organs in the region of pressure is evaluated, with an assessment of the degree and permanence or irreversibility of their damage. Imaging tests are repeated many times during the postoperative period and are also the basis for the clinical assessment of a patient in a long-term follow-up.

Echocardiography

Two-dimensional echocardiography (ECHO) provides excellent imaging in the youngest children, newborns and infants. Currently, this is a standard examination performed in newborns with heart and vessel anomalies. Very often it is sufficient for the proper eligibility for surgical treatment. It is also a basic postoperative follow-up test.

Angiography

Traditional angiography (ANGIO) is used in case of echocardiographic doubts or in patients with unclear aortic arch anatomy, those with suspicion of additional anomalies and in older patients. Angiographic examination allows a precise assessment of most arterial vessels and blood flows but requires the administration of contrast agents.

Magnetic resonance imaging

The aortic arch and proximal segment of the descending aorta are well represented structures in magnetic resonance imaging (MRI). This method provides better 2D images and spatial reconstructions than any other non-invasive technology. MRI also allows detailed imaging of the vasculature in relation to other mediastinal structures. In this examination it is also possible to evaluate the condition of tissues and organs adjacent to the vessels.

Computed tomography

The main advantage of computed tomography (angio-CT) is the precise imaging in patients after implantation of metal stents and with high risk of forming aneurysms in the postoperative period and with contraindications to use MRI [8].

Methods of functional evaluation of the patient’s upper extremities in the remote period after aortic arch vessel surgery

Prior to the anthropometric, biomechanical, and functional evaluation of the upper extremities, the functional hand
dominance should be determined. Information concerning right- or left-handedness is necessary to carry out a comparative analysis of extremities in terms of their efficiency or strength, especially in older children. Considering the obtained test results exclusively through the prism of the right and left side of the body may disrupt the process of deducing and be of no use to clinicians and patients, especially since the percentage of left-handed people is approximately 10% of the population [9]. One of the tools to determine the functional dominance of the upper limb in a child is the Peg Moving Task (PMT-5). The test consists in placing 5 sticks in the holes of a wooden board as soon as possible. The hand that completed the task in a shorter time is considered as dominant [10]. In older children, the dominant upper limb is the hand the child writes, uses a spoon, toothbrush, or scissors [11].

Examining the length and circumferences of the upper limb
Disturbance in the main arterial blood flow to the upper limb in adults usually results in the establishment of collateral circulatory system capable of maintaining limb life and function. In children, a similar dysfunction may adversely affect the nutrition of limb tissues, and thus disturb their growth and function [12]. In addition to measurement of the relative and absolute length of the upper limbs, it is useful to include segmental measurements of the arm and forearm to accurately determine the region of long bone growth disorder. Measurements of circumferences, particularly the first (R1) arm (short and long) one, second (R2) arm one and first (P1) forearm one, can provide information concerning soft tissue hypotrophy.

Hand grip strength (HGS) test
The Electro-Hydraulic Hand Dynamometer is acknowledged by the American Society of Hand Therapist as an objective tool for monitoring HGS in patients after cardiac surgery [13]. The standard protocol assumes the measurement of isometric force in a standing position [14], therefore, by reason of the test specificity and the possibility of a sudden increase in blood pressure, a sitting position with the arm slightly splayed from the body and the limb bent in the elbow joint up to 90° is recommended in cardiac patients (according to JAMAR protocol) [15]. The maximum hand grip must be maintained for at least 3 seconds [16]. The device is standardized and can be used in patients aged over 4 [17].

Muscle strength test
Biodex System PR04 Dynamometer allows precise measurement of muscle strength moments under isometric (static), isokinetic (dynamic) and isotonic conditions in both healthy people and those in the rehabilitation process. The device can be used to objectively measure the strength for individual limb movements in the humeral [18] and elbow joints [19] within the full range of patient movements [20]. In addition, it is possible to determine the relationship between antagonistic muscle strength. The aim is to determine the proportion of muscle strength within a given joint [21, 22]. The measurement of muscle strength moments for both upper limbs makes it possible to directly compare them and determine the strength symmetry [21].

In patients after cardiac surgery it is not recommended to perform measurements under isometric conditions due to the excessive strain on the body by static work. In the rehabilitation process, measurements are frequently performed under isokinetic conditions [22, 23], in which the movements are performed with a constant, programmed by the researcher, value of angular velocity in the joint, and the generated resistance is adjusted individually to the patient’s abilities.

The data used for the analysis is the maximum strength moment reached at a given angular velocity (absolute and relative values, i.e. related to the examined person’s body weight), total work, maximum and average strength and fatigue index. The value of the strength moment may be affected by such factors as age, gender, BMI, or extremity dominance [24]. It is possible to adapt the dynamometer arms for children aged 6 and older [25].

Testing of electrical activity of muscles
Electromyography (EMG) is concerned with the study of changes in the electrical activity of muscles in particular time periods [26]. Surface electromyography (sEMG) is useful in complementing the diagnosis of neuromuscular diseases in children [27]. Due to possible postoperative neurological complications in cardiac patients, conducting a non-invasive examination using sEMG while performing selected motor tasks may provide information concerning the existing disorder and differences in muscle activation patterns [26]. Commonly used recommendations for placing electrodes on the skin are SENIAM guidelines [28].

To compare the examined patients to one another and possibly monitor the rehabilitation process, it is necessary to normalize the EMG signal amplitude, i.e. to compare the measured value with a reference value, which is obtained depending on the selected method. The MVIC (MVC) technique (maximum voluntary isometric contraction) assumes the execution of maximum isometric contraction [26]. The validity of this normalization technique should be considered in the case of cardiac patients in whom this could lead to deterioration of their health status. Other ways of normalization include obtaining of a reference value from the test, taking the maximum amplitude value of 100% or an average value, and it is also possible to measure the reference value for the conditions of known, non-maximum constant loading of the muscle group under static conditions [26].

The analysed data usually include the mean amplitude of the signal, the surface area under the signal envelope, the
mean and median frequency spectrum of EMG signal and the analysis of temporal characteristics of the muscle function. Due to the ease of testing, electromyography has become a common tool to study the action potential of muscles, but it should not be equated with measuring muscle strength or power [26].

**Infrared thermography imaging**

The undisturbed function of the cardiovascular system is a precondition for appropriate development of the limb and maintaining its full functionality. The cardiovascular system is responsible i.a. for maintaining adequate temperature of tissues, thus infrared thermography imaging may be helpful in finding impaired blood supply in the upper limbs. They may manifest as a decrease in temperature of peripheral structures. Infrared thermography imaging, due to its minimal invasiveness and ease of use, is applied in body temperature tests in both adults and children [29]. This examination can only complement the basic tests, which assess blood flow in upper limb vessels. The existing literature does not contain clear indications for the use of infrared thermography and diagnostic protocols.

**Small motor skills test**

The assessment of the functional condition of the upper limb should be based on the small motor skills test. A commonly used tool is Assisting Hand Assessment (AHA). AHA measures and describes how children with one-sided disability use the affected hand to perform tasks/activities involving both hands. It consists of 22 parts subject to observation, which, depending on the quality of performance, are rated from 1 to 4 points [30]. AHA has been developed for children aged between 18 months to 12 years and allows monitoring of the rehabilitation process [31]. For children aged between 18 months and 5 years, the study involves observation of spontaneous play with toys requiring two hands (Small Kids AHA). A board game (School Kids AHA) has been developed for children aged 6–12 years, which provides an age-appropriate context for using the same toys as AHA for small children [32]. AHA is considered a useful clinical and research tool [33]. In 2017, the first tool assessing the hand function in children aged 3–12 months was developed — Hand Assessment for Infants (HAI). This tool consists of 31 parts assessed on a scale from 0 to 2 points depending on the quality of the presented infant’s motor skills [34]. Both AHA and HAI are used in the diagnosis of children with neurological disorders, especially with cerebral palsy [30, 34].

**Evaluation of psychomotor development of children after aortic arch surgery**

In addition to the above instruments, which are strictly dedicated to the assessment of upper limb functions, there are a number of tools for comprehensive evaluation of the child’s psychomotor development, the individual modules of which also cover the areas of small motor skills. In 2012, the American Heart Association (AHA) published a statement recommending continuous monitoring of the development of children and adolescents with congenital heart defects using standardized assessment tools. To determine the level of development of small motor skills and hand functions, AHA recommends using the following methods and tests: the Bayley Scales of Infant and Toddler Development — Third Edition, Bruininks-Oseretsky Test of Motor Proficiency — Second Edition (BOT-2), Grooved Peg Board, NEPSY-II, Peabody Developmental Gross Motor Scale (PDMS-II), Scales of Independent Behavior — Revised (SIB-R), Vineland Adaptive Behavior Scales — Third Edition (Vineland-3), Test of Visual-Motor Integration, Supplemental-Motor Coordination II (VMI) [35].

The Bayley Scales of Infant and Toddler Development (Bayley-III) is the “golden standard” in the assessment of infants and children with developmental disorders, as well as in monitoring the effectiveness of the therapeutic measures taken. The Bayley-III is a standardized tool for evaluating the development of children aged from 1 month to 42 months. The Bayley-III contains a subscale evaluating the development of small motor skills, i.e. precision hand movements. The subscale of precision motor skills is part of the overall assessment of a child’s development but can also be treated as an independent part of the test. The evaluation of small motor skills is based on the examination of 66 characteristics and measures such abilities as hand grip quality, manual dexterity, handling of objects, visual-motor coordination, and movement dynamics. The scores obtained in the test are compared with adopted standards [36].

The above evaluation tools are also useful to comprehensively assess the motor deficits of the upper limbs, which may occur after cardiac surgery in the youngest patients.

**Conclusions**

Anthropometric, biomechanical and functional evaluation of upper extremities in children in long-term follow-up after surgery of aortic arch vascular anomalies allows an additional evaluation of the effectiveness and possible side effects of individual surgical methods.

Monitoring the development and efficiency of upper extremities in children treated for the most common congenital heart and vascular defects, with consequences in the form of blood supply disorders, should be based on a basic hemodynamic evaluation, as well as repetitive, non-invasive diagnostic tools and tests that check structural lesions within the upper limbs, as well as function tests, including, above of all, small motor skills.
Streszczenie

Wrodzonym wadom serca często towarzyszą wady wielkich naczyń, takie jak koarktacja aorty z różnymi formami hipo- plazji łuku aorty, pierścienie naczyniowe. Leczenie operacyjne i wybór optymalnej techniki korekcji mogą być zaburzenia perfuzji, także w zakresie naczyń zaopatrujących kończyny górne, najczęściej lewą kończynę górna.

Słowa kluczowe: wrodzone wady serca, wady naczyń łuku aorty, kardiochirurgia dziecięca, koarktacja aorty, hipoplazja łuku aorty, korekcja kardiochirurgiczna, ocena kończyn górnych, siła mięśniowa, EMG, AHA

Folia Cardiologica 2020; 15, 5: 372–379

References


