

# High risk HLHS: hybrid approach yes, but how does it work?

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The majority of newborns with duct-dependent systemic blood flow are related to a hypoplastic left heart syndrome (HLHS), accounting for 2–3.5% of congenital heart defects (1). HLHS is anatomically subdivided in defects with stenosis or atresia of the mitral and aortic valve; consecutively left ventricular chamber size varies between fully diminished to almost normal dimensions in case of an associated VSD. Untreated, HLHS patients died in about 90% within the first week of life (1). Restrictive atrial septum is observed in about 22%; 6% of the HLHS newborns have an intact atrial septum and are critically diseased because of severe postnatal hypoxemia (2). Considering a balanced pulmonary to systemic circulation, clinical condition remains stable. Postnatal adaptation usually leads to a decrease in pulmonary vascular resistance; consecutive diastolic left-to-right shunt with increased oxygenated blood passing the duct disposed together with missing maternal prostaglandin for postnatal duct obstruction. Newborns with pre- and postnatally undetected duct-dependent systemic blood flow are oftentimes admitted in cardiogenic shock; pulmonary run off and/or duct obstruction are the most reasons. Preoperative hemodynamic instability, moderate-to-severe dysfunction of the dominant ventricle and atrioventricular valve regurgitation, restrictive atrial septum and/or obstructed total anomalous pulmonary venous return classifies HLHS-newborns as high risk (3); further birth weight less of 2.6 kg as well as prematurity (<37 weeks). Considering a Norwood surgery in high-risk HLHS patients, the Aristotle score increases from an already high baseline value of 15 to

19 or even above 20 calculated points; latest is associated with a dramatic decrease of the post-surgical survival rate (3,4). Exact 20 years ago, Hakan Akintuerk performed in Giessen, Germany the first successful comprehensive stage II in a 5-month-old HLHS patient (5). The patient admitted as 2-day-old newborn in a cardiogenic shock; bilateral pulmonary banding (bPAB) solved immediately the cause of pulmonary run-off; his duct-dependent circulation was initially treated by a continuous infusion of PGE1. Two days after bPAB the duct was stented as an elective percutaneous transcatheter approach. Currently, the 20-years old man still lives in a good clinical condition. Based on our initial experiences in high-risk HLHS neonates (5), later on Norwood surgery was replaced also as an elective procedure. Outcome data did not show any difference between high-risk and HLHS neonates with a normal risk score. Hybrid stage I can be performed with insignificant mortality (6,7). Expanded experiences, improvements of techniques, in particular stent material, nearly all HLHS newborns can currently have a chance for treatment. Additionally, hybrid approach was also introduced to transfer newborns with high-risk of non-HLHS from the neonatal period in late infancy to achieve either an option for biventricular repair or to reduce neonatal surgical risk (8,9). Shifting an extensive open-heart surgery beyond the neonatal period improved also the neurological development comparing conventional surgical procedures (10,11). However, aim and technique of hybrid approach consisting of bPAB, ductal stenting and manipulation of the atrial septum and aortic isthmus,

if necessary varied worldwide between Pediatric Heart Centers (6,12-15). In Giessen Germany, only bPAB is surgically performed as a component of the hybrid approach; any additional risk as transpulmonary duct stenting is avoided. Duct stenting and if necessary aortic arch manipulation and/or atrial septostomy are performed by a percutaneous transcatheter approach as an elective follow-up procedure (6,7). The “Hybrid” group in Columbus introduced transpulmonary duct stenting during the same open chest surgery, immediately after bPAB completion (12). In Toronto Canada (13), additionally a reverse shunt procedure was institutionalized with aim avoiding a supposed descending aortic arch obstruction compromising a sufficient retrograde blood flow to the cerebral and coronary circulation. Postnatal hybrid procedure is even used for bridging to a Norwood stage I beyond the neonatal period by combining bPAB with continuous infusion of prostaglandin E1 (14) or together with a transpulmonary duct stenting (15).

The report of Nwankwo *et al.* (16) “Hybrid Strategy for Neonates with Ductal-Dependent Systemic Circulation at High Risk for Norwood” underlines the need for an additional surgical option beyond the Norwood procedure treating newborns with duct-dependent systemic blood flow. It seems that a good prepared hybrid-program might be of benefit for many patients in any Pediatric Heart Center, independent from an out-standing Norwood program. The Pittsburgh study presents retrospective results based on a pre-selected institutional risk stratification management. The annual number of about 10 Norwood stage I procedures with a 30-day mortality of 8% or even less than 4% neglecting high risk patients represents a middle-sized Pediatric Heart Center with a high-leveled care of HLHS and variants. Before a hybrid program was established, 16 patients classified as high risk received a Norwood procedure. This advanced neonatal surgery was performed on expense of 31% mortality by neglecting further morbidity data. After an established hybrid-program, the thirty-day mortality of the high-risk neonates could be reduced to less than 4%. However, the interstage mortality and comprehensive stage II results equalized the initial benefits of hybrid stage I. Considering these data, it seems that there is still space for improvement. It remains open, if techniques of the hybrid stage I and/or interstage monitoring have the main impact on the final results. Based on similar experiences as achieved in Pittsburgh, many centers recommend the hybrid approach as an interim procedure to rescue high-risk patient independent if during

the follow-up a Norwood stage I surgery or comprehensive stage II approach is performed (15). Currently, the decision to adopt the hybrid approach or the Norwood approach seemed to be based on the preference of congenital heart surgeon’s rarely pediatric cardiologists. Over the last 2 decades, many institutional reports and reviews were published to figure out the “pros and cons” of hybrid approach in context of the “conventional” Norwood stage I surgery (17,18). Hybrid stage I together with its later performed comprehensive stage II was often compared to Norwood stage I with its necessitating follow-up bi-directional Glenn-shunt. Over all, the hybrid-approach was and is mostly in the defensive, apostrophized between bad and questionable or as the “fifty shades of grey” in comparison to Norwood procedure (19). Conclusions oftentimes ended with the summary of missing scientific studies, which could support the possible benefits of the hybrid approach (20). In this context, following questions arise: (I) why is an alternative to a standard therapy be battled, if nobody is obligated to change the routine? (II) Why do always find difficult lateral options for treating our patients? (III) How can be a surgical approach an option for “high”-risk, but not for elective patients? May be the last question should be answered first. Based on 20-year experience treating almost 250 patients by a hybrid approach in setting of HLHS or newborns with duct dependent systemic blood flow with borderline left heart structures, my subjective answer is, that there is not a shame for any Pediatric Heart Center having an additional hybrid—despite a well-going Norwood-program. However, that implies like in Pittsburgh, surgeons and cardiologists have to be prepared for performing and establishing a hybrid approach. The beginner team has not to start, as we 20-year ago, just confronted to a HLHS newborn in cardiogenic shock. The easy looking three hybrid parts—bPAB, duct stenting and atrial septum/isthmus manipulation—need training and appropriate material and surgical and interventional skills resulting in acceptable results. Surgically performed bPAB can be a disaster or a benefit for the necessitating follow-up operations ending in a hemodynamically well functional Fontan circulation. The PAB can be associated with hypoplastic branches or if appropriate performed with good developed pulmonary arteries based on a favored, post-(stenotic) band pulmonary artery dilatation. For this the PAB width should be small with a diameter of 1–2 mm, fixed at the adventitia, and depending of the body weight with a diameter of 3.5 or 3 mm (6). Considering that duct-stenting is not a must (14),

stent placement should be performed as transpulmonary or as a separate, elective percutaneous approach. We prefer duct stenting as a separate approach with self-expandable stent-material, which has in Europe a CE-mark for newborns with duct dependent-systemic blood flow (6). One of the main reasons for percutaneous duct stenting is the direct impact on the junction of the duct to the descending aortic arch. The isthmus region needs sometimes be manipulated both, before or after stent is positioned within the duct. Important is in any case having a favorable isthmus region. If not, the aortic isthmus might be balloon dilated before duct is stented or an aortic coarctation becomes evident following placement of a duct stent, then an additional stent is necessary treating the coarctation. Again, also the Pittsburgh experience shows, that the interstage remains a vulnerable period until the comprehensive stage II is completed. The strategy to utilize the hybrid approach for bridging to Norwood-stage I and not to a comprehensive stage II might be born on similar problems during the hybrid-interstage (15).

In conclusion, why hybrid? (I) Each of the three parts of the hybrid approach can be utilized as one of the most effective resuscitative tools for newborns with duct-dependent systemic blood flow; independent of the cause of the compromised cardiovascular function; (II) hybrid approach generates chances for high-risk HLHS and duct-dependent non-HLHS newborns; (III) hybrid approach is able to replace neonatal Norwood stage I surgery; (IV) hybrid approach is and must remain the minimal invasive alternative of the Norwood surgery. Considering the Editorial of Charles Fraser to the surgical palliation for HLHS “for now, just keep doing what you do best” (21), it can be expanded to the hybrid approach “looks simple, requires extensive training” (22).

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## Footnote

*Conflicts of Interest:* The author has no conflicts of interest to declare.

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