The Journal of Maternal-Fetal & Neonatal Medicine

ISSN: 1476-7058 (Print) 1476-4954 (Online) Journal homepage: www.tandfonline.com/journals/ijmf20

Pregnancy in ACHD women: crucial role of multidisciplinary clinical roadmap

M. Grandinetti, S. Salvi, A. Olimpieri, S. Fruci, E. Portinaro, K. Corigliano, R. Lillo, M.C. Meucci, F. Graziani, A.B. Delogu, M.L. Narducci, S. De Carolis, G. Vento, D. Arduini, A. Amodeo, A. Lanzone & M. Massetti

To cite this article: M. Grandinetti, S. Salvi, A. Olimpieri, S. Fruci, E. Portinaro, K. Corigliano, R. Lillo, M.C. Meucci, F. Graziani, A.B. Delogu, M.L. Narducci, S. De Carolis, G. Vento, D. Arduini, A. Amodeo, A. Lanzone & M. Massetti (2025) Pregnancy in ACHD women: crucial role of multidisciplinary clinical roadmap, The Journal of Maternal-Fetal & Neonatal Medicine, 38:1, 2470411, DOI: 10.1080/14767058.2025.2470411

To link to this article: https://doi.org/10.1080/14767058.2025.2470411

© 2025 The Author(s). Published by Informa UK Limited, trading as Taylor & Francis Group

Published online: 10 Mar 2025.

Submit your article to this journal

Article views: 1870

View related articles

View Crossmark data

Citing articles: 1 View citing articles

Full Terms & Conditions of access and use can be found at
https://www.tandfonline.com/action/journalInformation?journalCode=ijmf20

THE JOURNAL OF MATERNAL-FETAL & NEONATAL MEDICINE
2025, VOL. 38, NO. 1, 2470411
https://doi.org/10.1080/14767058.2025.2470411

ORIGINAL ARTICLE

OPEN ACCESS

Pregnancy in ACHD women: crucial role of multidisciplinary clinical roadmap

M. Grandinetti[a,b**], S. Salvi[c,d**], A. Olimpieri[a,b], S. Fruci[c,d], E. Portinaro[a,b], K. Corigliano[a], R. Lillo[a,b], M.C. Meucci[a,b], F. Graziani[a,b], A.B. Delogu[e,f], M.L. Narducci[a,b], S. De Carolis[c,d], G. Vento[e,f,g], D. Arduini[g], A. Amodeo[a,b,g], A. Lanzone[c,g] and M. Massetti[a,b,g]

[a]Dipartimento di Scienze Cardiovascolari, Fondazione Policlinico Universitario A. Gemelli IRCCS, Roma; [b]Dipartimento di Scienza cardiovascolari e Polmonari, Università Cattolica del Sacro Cuore, Roma; [c]UOC di Ostetricia e Patologia Ostetrica, del bambino e di sanità Pubblica, Fondazione Policlinico Universitario A. Gemelli IRCCS, Roma; [d]Dipartimento Universitario di Scienze della Vita e Sanità pubblica, sezione di Ginecologia ed ostetricia, Università Cattolica del Sacro Cuore, Roma; [e]UOC di Pediatria, del bambino e di sanità Pubblica, Fondazione Policlinico Universitario A. Gemelli IRCCS, Roma; [f]Dipartimento di Pediatria, Università Cattolica del Sacro Cuore, Roma; [g]Università Cattolica del Sacro Cuore, Roma

ABSTRACT

Purpose: Pregnancy in women with adult congenital heart disease (ACHD), whether in its natural history or after surgical correction, represents a unique pathophysiological model that requires careful, multidisciplinary management to ensure favorable maternal, fetal and neonatal outcomes. Investigating the impact of congenital cardiac conditions on maternal and feto-neonatal health, the effect of pregnancy-related cardiovascular changes on maternal cardiac health, and the outcomes for offspring born from ACHD mothers is highly relevant, due to the increasing number of ACHD women reaching adulthood and the significant burden these pregnancies can pose. The aim of this article is to provide food for thought to those who have always been involved in ACHD and pregnancy, but also to provide a training tool for young doctors who are approaching at this wonderful world for the first time.
Materials and methods: This article was conceived and structured as an "educational and debate". In this article we describe our experience in the ACHD outpatient clinic and the High-Risk Pregnancies Division of Fondazione Policlinico A. Gemelli Hospital IRCCS from 2013 and now includes over 100 patients evaluated over a 10-year period.
Results: In this article we describe our clinical pathway and the clinical history of our first patient, a 30-year-old woman with univentricular heart (criss-cross heart, double outlet right ventricle and pulmonary stenosis) who underwent a Glenn operation as a child. Our plan included scheduled cardiological and obstetrical follow-ups, as well as planned hospitalizations. An elective C-section was carried out at 38 gestational weeks under spinal anesthesia, with Extracorporeal Membrane Oxygenation and the heart surgery team stand by. It was an uncomplicated delivery. As a result, we developed a specific clinical pathway named "ACHD Pregnancy Pink Pathway."
Conclusions: The strength of this idea dwells in the synergy between different experts in deciding for the best decision regarding the required monitoring strictness and the more appropriate obstetric surveillance and delivery plan for the patient. The lesson we learned over the years is that to ensure the best diagnosis and treatment for our young unique patients, we must create a detailed "ROADMAP" for them. We propose a pioneering pathway divided into the three essential phases: maternal, obstetrics and fetal-neonatal.

ARTICLE HISTORY
Received 23 October 2024
Revised 5 February 2025
Accepted 15 February 2025

KEYWORDS
Pregnancy; ACHD; multidisciplinary; obstetrics; cardiovascular

Education and debate

This article was conceived and structured as an "educational and debate" with the aim of providing food for thought to those who have always been involved in ACHD and pregnancy, but also to provide a training tool for young doctors who are approaching at this wonderful world for the first time.

Pregnancy is a resource-demanding period for a woman's entire body, during which it undergoes numerous physiological changes to support the normal pregnancy progression, to balance the fetus's metabolic requests and maintain mother's homeostasis. Thanks to advancements in pediatric cardiac surgery, we are witnessing an increasing number of female

CONTACT E. Portinaro elena.portinaro@gmail.com Dipartimento di Scienze Cardiovascolari, Fondazione Policlinico Universitario A. Gemelli IRCCS, Roma; Dipartimento di Scienza cardiovascolari e Polmonari, Università Cattolica del Sacro Cuore, Largo A. Gemelli, 1, RM 00168, Roma
*These authors contributed equally to this paper.
This article has been republished with minor changes. These changes do not impact the academic content of the article.
© 2025 The Author(s). Published by Informa UK Limited, trading as Taylor & Francis Group
This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0/), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited. The terms on which this article has been published allow the posting of the Accepted Manuscript in a repository by the author(s) or with their consent.

grown-ups' patients, born with different congenital heart diseases, who get pregnant. Care of these ACHD patients represents a new major challenge, and the establishment of dedicated centers has been proven to improve outcomes in this patient population.

One of the challenges in this population is the control of risk factors as risks to the mother, fetus, and offspring all coexist. The risk of cardiovascular events, such as of arrhythmias, cyanosis, pulmonary hypertension and heart failure, in ACHD pregnant women is primarily related to the specific congenital condition, previous surgeries, and the current maternal cardiac status. Different authors have developed different score systems, such as the Cardiac Disease in Pregnancy [1] Risk Score and the ZAHARA Score [1,2]. Risk stratification for cardiovascular diseases in pregnancy divides patients into four groups based on disease severity, according to the Modified World Health Organization Classification of Maternal Cardiovascular Risk (mWHO) [3]. Women in the moderate and high-risk groups require thorough pre-conceptional evaluation, care in a dedicated expert center for pregnancy management, and organized delivery planning.

Previous studies have emphasized that cardiovascular diseases in pregnancy are significant risk factors for both obstetric outcomes (with Caesarean-section rates ranging from 36% to 60% depending on the severity of cardiovascular disease, compared to the overall reported of 26% in Europe) [4] and cardiological outcomes (such as heart failure, occurring in 5% of cases in mWHO I and 33% of cases in mWHO IV) [3].

Furthermore, ACHD women's fetuses and newborns have an increased risk of unfavorable outcomes for what concerns: natural or iatrogenic prematurity (12–28%), intrauterine death (3%); neonatal mortality in the first week of life [5,6].

There is a lack of studies concerning the outcomes for fetuses and newborns carried by ACHD women. Therefore, it is important to investigate strategies for preserving the health of mothers, fetuses and newborns, given that WHO classification is directly related to perinatal complications and prematurity.

Our experience began in 2013 and now includes over 100 patients evaluated over a 10-year period in the ACHD outpatient clinic and the High-Risk Pregnancies Division of Fondazione Policlinico A. Gemelli Hospital IRCCS.

The first patient in our Clinical Pathway is a 30-year-old woman with univentricular heart (criss-cross heart, double outlet right ventricle and pulmonary stenosis) who underwent a Glenn operation as a child. She had previously been lost to follow-up and has moderate functional capacity. She was in her fourth pregnancy with a history of four previous miscarriages. At 10 weeks of gestation, the clinical assessment revealed impaired oxygen saturation (SpO2) of 85% on room air and digital clubbing. Maternal echocardiography showed preserved function of the single ventricle but severe atrioventricular valve regurgitation, placing her at high risk for heart failure.

The clinical case was discussed by our multidisciplinary ACHD pregnancy Team. The high risk of maternal morbidity and mortality along with fetal and neonatal complication, was discussed with the patient. A unique personalized protocol for pregnant women with univentricular hearts was then developed. In particular, a two-week cardiac assessment was established to identify signs of cyanosis early. This protocol included maternal cardiac monitoring every two weeks, a complete blood count every three weeks to manage polycythemia, and recommendations for bed rest. Maternal obstetric evaluation and fetal ultrasound surveillance were scheduled every four weeks.

The pregnancy was uneventful. The second trimester scan revelead no fetal anomalies and no cardiac malformations were detected with fetal echocardiography; fetal growth was normal with the last scan revealing a fetal weight estimated at 35° pc. The woman remained asymptomatic for cardiac warning signs. She was followed in the outpatient clinic for high risk pregnancies in conjunction with cardiologists and cardiac surgeries; at 37 weeks admission was planned to determine the timing and mode of delivery (Figure 1). An elective C-section was then carried out at 38 gestational weeks under spinal anesthesia, with Extracorporeal Membrane Oxygenation and the heart surgery team stand by in a hybrid surgery room. It was an uncomplicated delivery. The woman was admitted to the intensive care unit for 24 h and experienced no complications. The neonatal Apgar was 9/10 and the neonatal hospital course was regular. They were both discharged 8 days after delivery with no complications. At the two months follow-up after delivery, there were no limiting symptoms during daily activity for the mum.

This case was unique in the literature. Although various scores exist for pregnancy risk stratification, the ACHD population is too heterogeneous to fit neatly into any of these categories. Therefore, clinical pathways should be tailored to each patient. This case demonstrated the necessity of a multidisciplinary approach to minimize pregnancy-related maternal and fetal risks and optimize long-term outcomes. As a result, we developed a specific clinical pathway named "ACHD Pregnancy Pink Pathway." During regular follow-up for congenital cardiac condition (in natural history, surgically corrected or palliated) patients underwent integrated checks by a multidisciplinary team including congenital cardiac surgeons, cardiologists, obstetrics and anesthesiologists. Timing of controls, mode of delivery and type of anesthesia have been discussed and "tailored" for each woman (Figure 2).

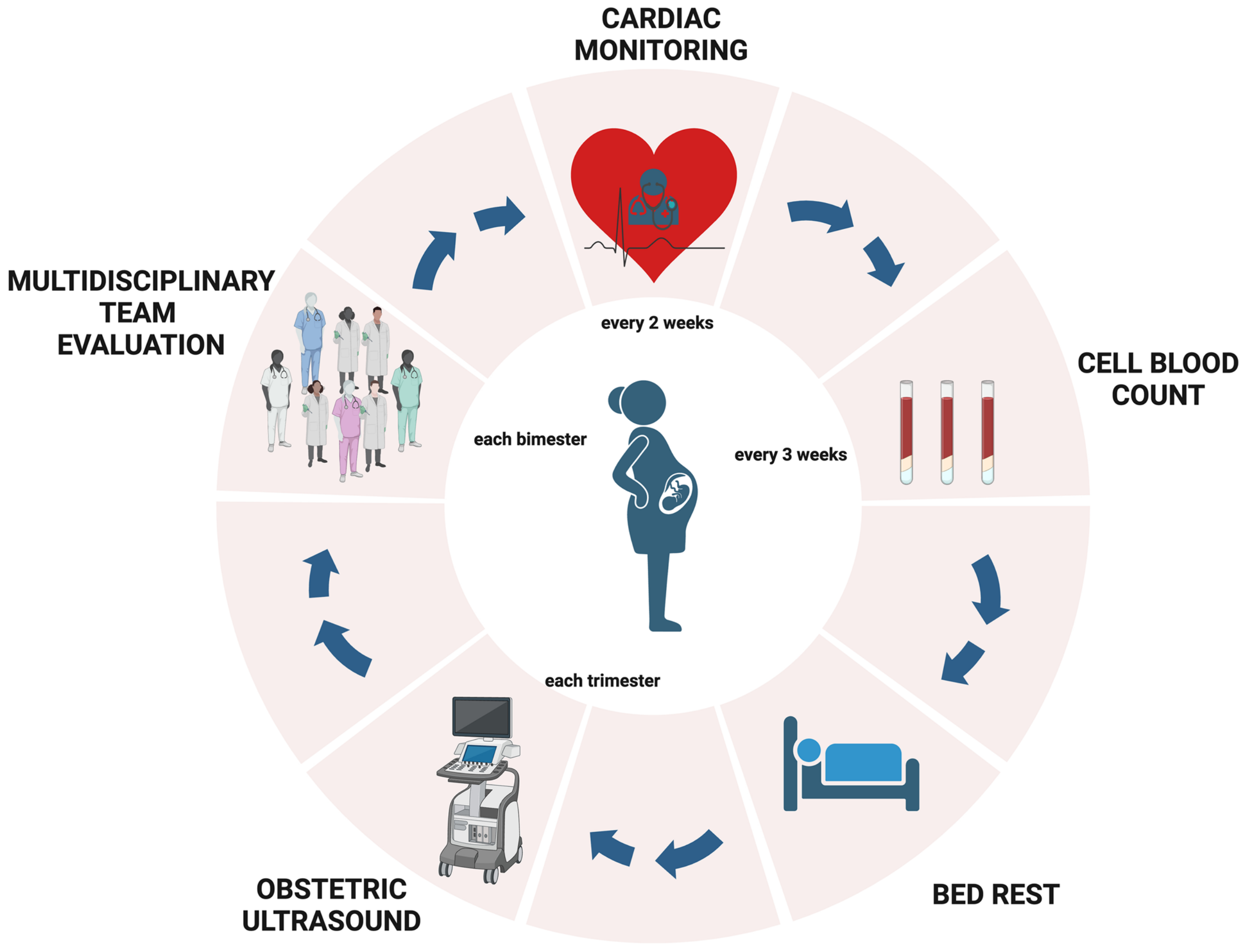

Figure 1. Our personalized protocol for pregnant women with univentricular hearts.

	CLASS I mWHO	CLASS II mWHO	CLASS II-III mWHO	CLASS III mWHO	CLASS IV mWHO
Preconception counseling	✓	✓	✓	✓	! Due to the highest maternal and feto-neonatal risks, pregnancy is considered contraindicated, necessitating accurate and comprehensive pre-pregnancy counseling !
I Trimester maternal cardiological and obstetric evaluation	✓	✓	TWICE	AT LEAST TWICE	MONTHLY
II Trimester maternal cardiological and obstetric evaluation	No if stable	✓	TWICE	AT LEAST TWICE	MONTHLY
III Trimester maternal cardiological and obstetric evaluation	No if stable	✓	TWICE	AT LEAST TWICE	MONTHLY

Figure 2. Our integrated flow-chart with integrated checks by a multidisciplinary team: timing of controls, mode of delivery and type of anesthesia have been discussed and "tailored" for each woman.

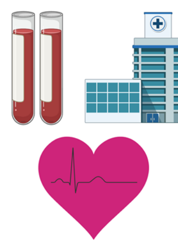

Figure 3. Our preliminary ACHD pregnancy pathway with three essential phases: maternal, obstetrics and fetal-neonatal. Abbreviations. ECG, electrocardiogram; Cardiopulmonary exercise testing; MRI, Magnetic Resonance Imaging; CT scan, computed tomography scan; CHD, congenital heart disease.

Our integrated clinical multi-flowchart was divided into the three essential phases: maternal, obstetrics and fetal-neonatal (Figure 3).

We proposed a pioneering pathway for ACHD women who are planning to become pregnant or who already are pregnant. A fundamental aspect of this approach is counseling. By implementing a tailored upfront plan before pregnancy and during the pre-conceptional period, we can establish a doable roadmap for these patients. This plan includes scheduled cardiological and obstetrical follow-ups, as well as planned hospitalizations.

The strength of this idea dwells in the synergy between different experts in deciding for the best decision regarding the required monitoring strictness and the more appropriate obstetric surveillance and delivery plan for the patient.

The lesson learned over the years is that to ensure the best diagnosis and treatment for our young unique patients, we must create a detailed *"ROADMAP"* for them. In a dedicated pathway all specialists (cardiologist, pediatric cardiac surgeons, obstetricians, anesthesiologists and more) must collaborate. In a kinky vision, comparing our job to contemporary financial world, we are the stakeholders with various milestones very different from each other: our patients, as milestones, are the most heterogeneous in cardiological clinical panorama. Therefore, a tailored clinical roadmap for each patient is essential in daily practice. The role of a dedicated clinical pathway within a safe environment, supported by a super-specialist and integrated approach, is crucial.

Disclosure statement

No potential conflict of interest was reported by the author(s).

Funding

The author(s) reported there is no funding associated with the work featured in this article.

References

[1] Siu SC, Sermer M, Colman JM, et al. Prospective multicenter study of pregnancy outcomes in women with heart disease. Circulation. 2001;104(5):515–521. doi: 10.1161/hc3001.093437.

[2] Drenthen W, Boersma E, Balci A, et al. Predictors of pregnancy complications in women with congenital heart disease. Eur Heart J. 2010;31(17):2124–2132. doi: 10.1093/eurheartj/ehq200.

[3] Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, et al. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy. Eur Heart J. 2018;39(34):3165–3241. doi: 10.1093/eurheartj/ehy340.

[4] European R. PERINATAL HEALTH to 2019.Core indicators of the health and care of pregnant women and babies in Europe from; 2015.

[5] Roos-Hesselink J, Baris L, Johnson M, et al. Pregnancy outcomes in women with cardiovascular disease: evolving trends over 10 years in the ESC Registry Of Pregnancy and Cardiac disease (ROPAC). Eur Heart J. 2019;40(47):3848–3855. doi: 10.1093/eurheartj/ehz136.

[6] Gelson E, Curry R, Gatzoulis MA, et al. Effect of maternal heart disease on fetal growth. Obstet Gynecol. 2011;117(4):886–891. doi: 10.1097/AOG.0b013e31820cab69.