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Review

From “not well seen” to “likely normal”: Basic echocardiographic imaging of the coronary arteries in pediatric patients for diagnosis of anomalous aortic origin

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ABSTRACT

Transthoracic echocardiography is the initial imaging modality used to screen for congenital coronary anomalies in the pediatric population. Successful imaging of the coronary arteries depends on multiple factors including imaging technique, patient body habitus, acoustic imaging window availability, and patient cooperation. A standardized transthoracic imaging protocol performed by an experienced echocardiographer has been shown to improve detection rates of anomalous aortic origin of the coronary artery. This paper reviews basic transthoracic echocardiographic assessment of the coronary arteries and techniques for image optimization. A thorough evaluation includes determining the originating sinus, proximal course, visualization of flow within the coronary arteries, and assessment of flow with color Doppler. Cine clips of two-dimensional and color Doppler imaging should be recorded. Our institution prefers using the color compare feature of the equipment and color cine images. Still frame images of the coronary arteries are not recommended since false dropout may cause an anomalous coronary artery to appear normal. We recommend interrogating the inter-arterial space between the aorta and the pulmonary artery in parasternal short-axis view with low velocity color Doppler to identify linear diastolic color flow pattern within the aortic wall. Acute angle takeoff in addition to the color flow within proximal coronary lumen can be used to make the diagnosis of a proximal intramural course of anomalous coronary artery. Imaging should also focus on demonstrating ostial origin from the aortic sinus and branching of the left coronary artery into left circumflex artery and left descending artery.

1. Introduction

Transthoracic echocardiography is the initial imaging modality used to evaluate for congenital coronary anomalies, the second most common cause of sudden death in young athletes in the United States [1]. Echocardiography allows for a rapid and noninvasive assessment, which can be done without fluoroscopy or contrast. Unfortunately, during many pediatric echocardiograms the coronaries are not well seen or incompletely evaluated. Successful imaging of the coronaries depends on a multitude of factors, including imaging technique, patient body habitus, acoustic imaging window availability, as well as patient cooperation during the study. Often these limitations may be overcome with image optimization and imaging technique. However, some limitations are inherent to echocardiography. Transthoracic echocardiogram has limited spatial resolution, making it difficult to distinguish high-risk characteristics of anomalous coronary arteries, such as intra-arterial and intramural courses [2]. False dropout may also cause an anomalous coronary artery from the inappropriate sinus to appear normal, especially if the coronary artery takes an intramural course and exits the aortic wall at the level of the appropriate sinus [3]. A standardized transthoracic imaging protocol performed by an experienced echocardiographer has been shown to improve detection rates of anomalous aortic origins of the coronary artery from the inappropriate sinus of Valsalva [2]. This review aims to discuss a basic standardized approach to evaluating the coronary arteries and techniques for image optimization. We hope this may be a useful and concise reference for basic coronary artery imaging. Many papers have been published reviewing the evaluation of anomalous coronary origins using echocardiography and advanced imaging modalities, and we recommend reviewing these references for further understanding of anomalous origin of the coronary arteries [2–5].

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Table 1
Summary of our recommended coronary preset settings. Adjustment of some of these settings may be needed to optimize the imaging for each patient.

Probe	Highest frequency suitable for adequate penetration
Color Scale	20–30 cm/s
Sector width	Small pie shaped 
Frame rate	>30 Hz (reduce two dimensional and color sector width if drops below 25 Hz)
Compression	35–40
Two dimensional gain	50%–60%
Persistence	Medium to high
Color flow optimization	High

2. Recommended coronary imaging settings

We can expect that coronary imaging will be part of a complete echocardiogram protocol, and will require adjusting the machine settings during the study to optimize coronary imaging. We recommend the echocardiographer follow standard image optimization techniques and use settings optimal for coronary imaging (Table 1). The highest frequency probe suitable for each patient should be used to optimize resolution. The color scale should be reduced to 20–30 cm/s during color Doppler evaluation to better visualize the flow within the lumen of the coronary artery. In addition, a small pie shaped sector should be utilized. The sector width should be just large enough to cover the proximal aortic sinus of origin, and proximal course of the coronary artery being imaged. Higher frame rates should be used, preferably above 30 Hz. The two dimensional and color sector width may be reduced to improve the frame rate if it drops below 25 Hz. Compression should be in the low range (35–40) for optimal differentiation of two-dimensional structures with more black-white imaging. Two dimensional and color persistence should be increased to medium or high, to allow for better color Doppler filling of the coronary artery lumen. The flow optimization setting should be set to high during color Doppler evaluation to highlight the flow within the coronary artery lumen.

3. Echocardiographic assessment

A thorough evaluation of the coronary arteries includes determining the sinus of origin, the proximal course, visualization of flow within the coronary arteries, assessment of the directionality of flow, and the level of origin ("takeoff") from the aorta (Table 2). In some disease states, measurement of the coronary artery diameter may also be indicated, in which case it should be measured from inner edge to inner edge. Cine clips of two-dimensional and color Doppler imaging should be recorded. Our institution prefers this to be done using the color compare feature of the equipment, which demonstrates a two dimensional image, as well as a color cine image of the coronary arteries. Still frame images of the coronaries are not recommended because false dropout may cause an anomalous coronary to appear normal. [2] We recommend interrogating the inter-arterial space between the aorta and the pulmonary artery in parasternal short-axis view with color Doppler to identify linear diastolic color flow pattern within the aortic wall. Acute angle takeoff in addition to the color flow within proximal coronary lumen can be used to make the diagnosis of a proximal intramural course of anomalous coronary artery [5].

4. Sinus of origin and proximal course

The evaluation should begin with determining the sinus of origin using two-dimensional and color Doppler. Two dimensional imaging alone cannot confirm the origin due to the potential for false dropout. The origins are best visualized in the parasternal short axis view. The right coronary artery should normally arise from the right sinus of Valsalva, at approximately the 11 o'clock position (Fig. 1). It is often better visualized with a slight counterclockwise rotation of the probe from the standard parasternal short axis imaging plane. In some instances, the echocardiographer may need to move the transducer slightly towards the patient's right and tilt the transducer either downwards or upwards to visualize the level of origin. This is often the case when the level of origin of the right coronary artery is different than the level of origin of the left. The normal left coronary artery should arise from the left sinus of Valsalva, at approximately the 4 o'clock position (Fig. 2). The left coronary artery may be better visualized with clockwise rotation of the probe from the standard parasternal short axis window. Slight movement of the transducer to the patient's left may also allow for evaluation of its bifurcation into the left anterior descending and circumflex arteries.

5. Normal and abnormal flow patterns

Color Doppler interrogation should also be done in the parasternal short axis view, to assess the directionality of flow within the coronaries and to assess for any filling defects. A right coronary artery with a normal origin from the right sinus of Valsalva should have a red color Doppler pattern, as the blood flows from the aorta and into the proximal right coronary artery, towards the transducer. A left coronary artery with a normal origin from the left sinus of Valsalva should also have a red color Doppler pattern, as the blood flows from the aorta and into the left main coronary artery, towards the transducer. Flow in the left anterior descending branch should also appear red as it courses towards the transducer in this image. Flow in the left circumflex branch may appear red or blue depending on its course and angle of imaging. Lack of flow in the coronary arteries should raise concern for ostial atresia or stenosis. Retrograde filling of the right coronary artery should prompt further evaluation for abnormal right ventricle to coronary connections, such as myocardial sinusoids. This is demonstrated by using color Doppler mapping with low Nyquist limit specifically over

Table 2
Summary of our echocardiographic interrogation protocol for complete coronary artery assessment. All recommended interrogation views may not be available in some patients, depending on their acoustic imaging windows.

Interrogation protocol	Right coronary	Left coronary
Origin	Parasternal short axis: Two-dimensional/color Doppler	Parasternal short axis: Two-dimensional/color Doppler
Assess proximal course	Parasternal short axis: Two-dimensional/color Doppler	Left main and bifurcation into left anterior descending and left circumflex: Parasternal Short axis: Two-dimensional/color Doppler Left anterior descending: Parasternal Long axis: Two-Dimensional/color Doppler
Assess level of origin/take off	Parasternal long axis: Two-dimensional/color Doppler	Apical 5 chamber: Two-dimensional/color Doppler Subcostal left anterior oblique: Two-dimensional/color Doppler
Directionality of flow	Parasternal short axis – color Doppler	Parasternal short axis – color Doppler
Assess for filling defects	Parasternal short axis - 2D/color Doppler	Parasternal short axis - 2D/color Doppler
Measure diameter (if indicated)	Parasternal short axis: Two-dimensional	Parasternal short axis: Two-dimensional (inner edge to inner edge)

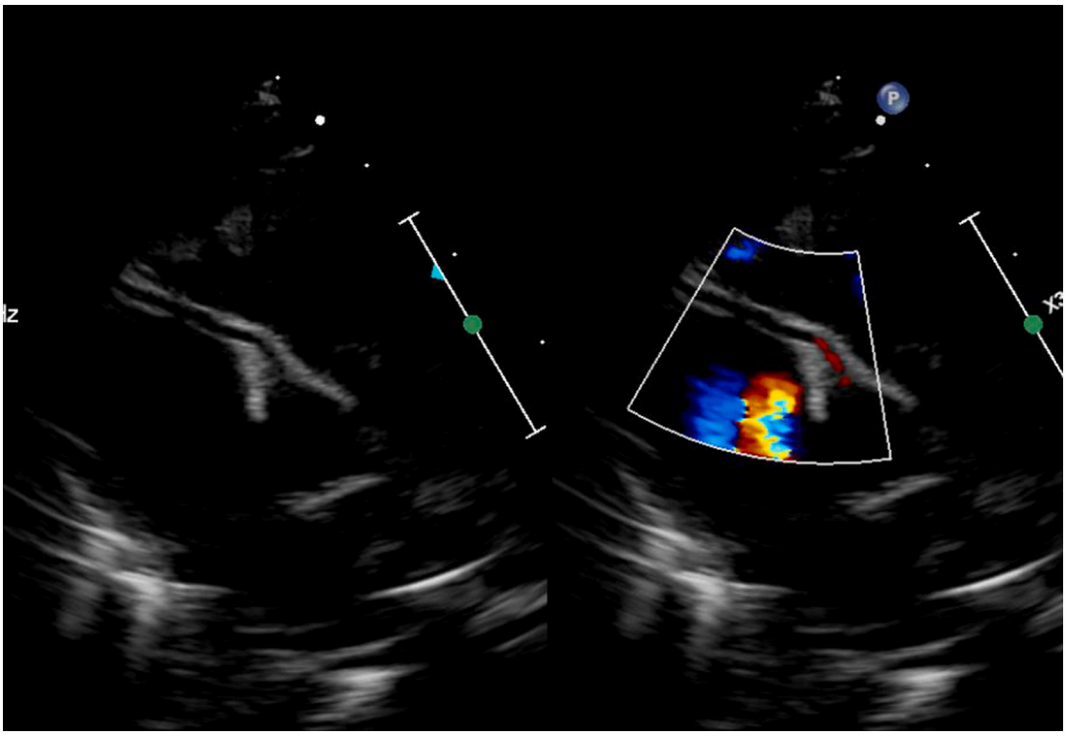

Fig. 1. Normal origin of the right coronary artery. In this picture the right coronary artery can be seen arising from the right sinus of Valsalva at the 11 o'clock position. Color Doppler imaging shows a red Doppler color pattern within the right coronary artery indicating appropriate flow from the aorta into the right coronary artery.

the interventricular septum, right ventricle, as well as the left ventricle, to assess for abnormal flow within the myocardium and along the endocardial surface. Retrograde filling of the left coronary artery should prompt concern for a possible anomalous origin, and is an essential echocardiographic component in the diagnosis of an anomalous left coronary artery origin from the pulmonary artery [3]. Coronary artery

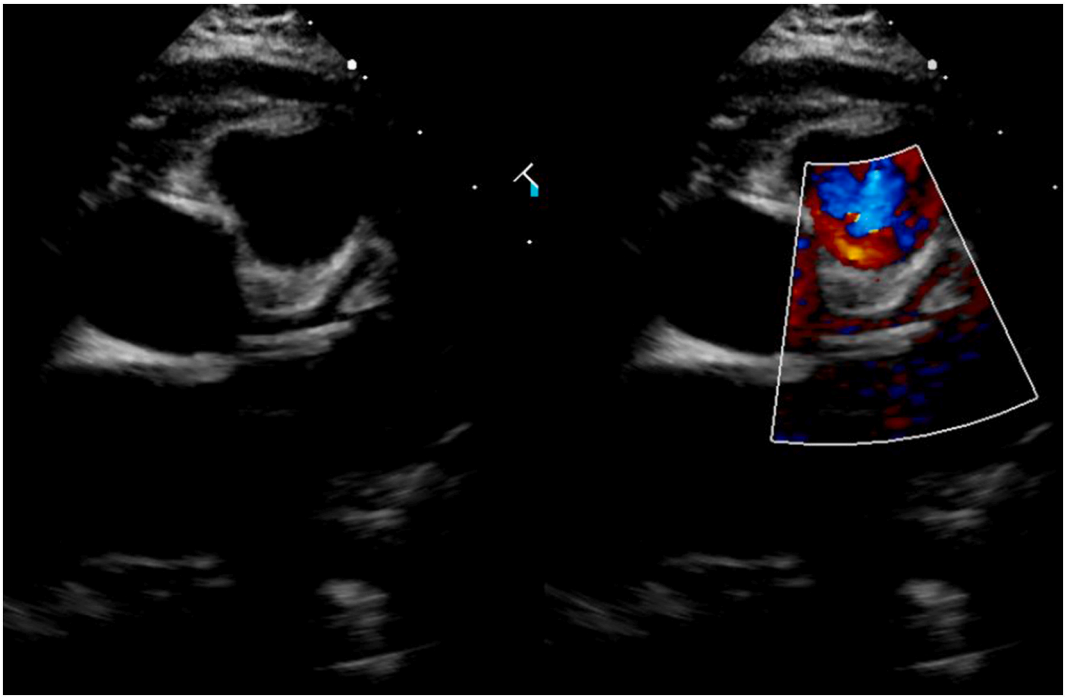

Fig. 2. Normal origin of the left coronary artery. In this picture the left coronary artery can be seen arising from the left sinus of Valsalva, at approximately the 4 o'clock position. Color Doppler imaging shows a red Doppler color pattern, indicating appropriate flow from the aorta into the left main coronary artery. The left main coronary artery can also be seen bifurcating into the left anterior descending and the circumflex coronary arteries. Flow within the normal left anterior descending branch should appear red as it courses upwards towards the transducer in this image. Flow within the normal left circumflex branch may appear red or blue depending on its course and the angle of imaging.

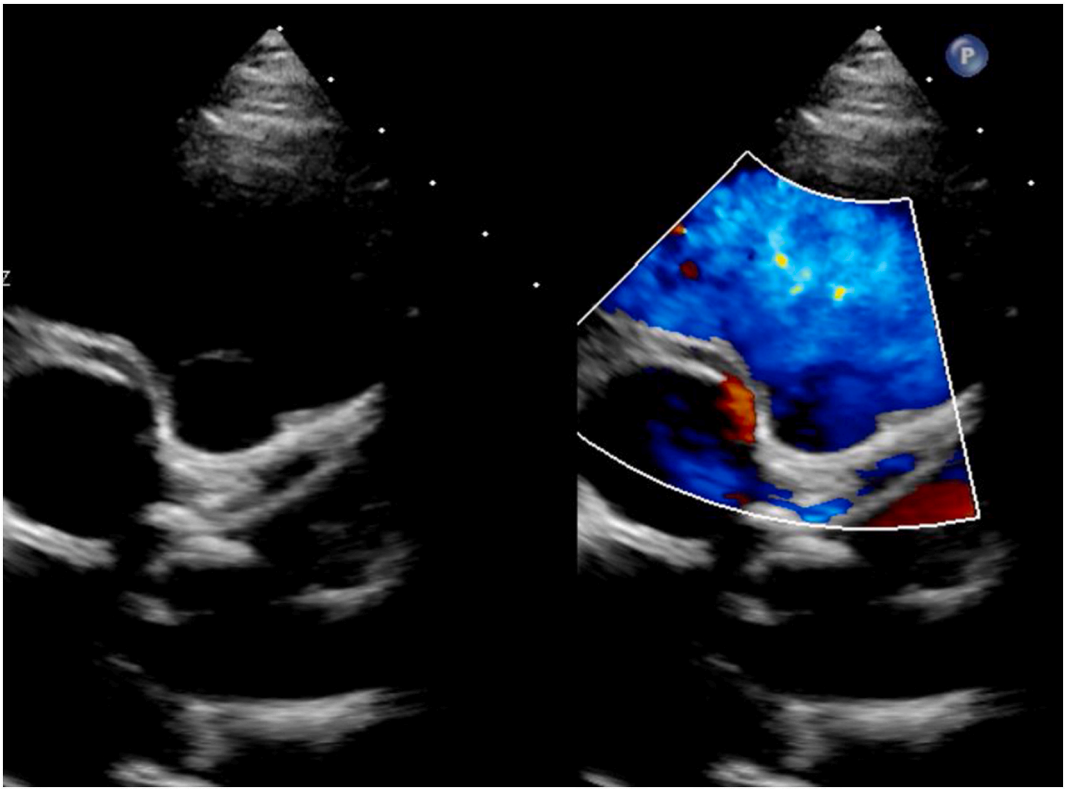

Fig. 3. Anomalous aortic origin of the right coronary artery from the left sinus of Valsalva, with an intra-arterial and intramural course. A red linear Doppler flow pattern is seen along the aortic wall consistent with an intra-arterial and intramural course. This diagnosis was confirmed with a coronary computed tomographic angiography (CTA).

fistulas may also be identified with color Doppler mapping. When draining into a low-pressure system, the fistula will be seen as a continuous high velocity flow, and when draining into a high- pressure system, will be visualized only in diastole. Dilation of the coronary arteries may be related to abnormal coronary connections such as fistulas, sinusoids, or Kawasaki disease. The proximal and well-visualized distal coronaries should be measured whenever dilation is suspected. An intra-arterial thrombus within the coronaries may be seen as an echogenic, two-dimensional structure within the coronary artery lumen. If a filling defect by Doppler imaging is present, it should prompt further evaluation with advanced imaging.

The intra-arterial space between the aorta and the pulmonary artery should also be interrogated by color Doppler, to rule out an anomalous coronary artery origin, specifically one with an intra-arterial course or intramural course. An anomalous coronary artery arising from the inappropriate sinus of Valsalva, with an intra-arterial and intramural course, may be identified on a transthoracic echocardiogram in the parasternal short axis view. The classic echocardiographic features are an acute angle of origin with a linear Doppler flow pattern along the aortic wall (Fig. 3). A red linear Doppler pattern is consistent with anomalous aortic origin of the right coronary from the left sinus of Valsalva, as the flow within the anomalous right coronary artery would appear red, as it moves upward and towards the transducer in this view. A blue linear Doppler pattern is consistent with anomalous aortic origin of the left coronary from the right sinus of Valsalva, as the flow within the anomalous left coronary artery would appear blue as it moves downward and away from the transducer in this view [5].

6. Level of origin

The level of origin or the “take-off” of the right coronary artery may be seen in the parasternal long axis view, with slight angling of the transducer to the patient's right (Fig. 4). This view should primarily be used to assess for a high level of take-off, at or above the level of the sinotubular junction, or from the ascending aorta. The echocardiographer may also use this view to better visualize the right coronary artery origin in the parasternal short axis view, by first identifying the level of origin in long axis and then carefully rotating into the parasternal short axis view. The parasternal long axis view should not be used in isolation to assess for a normal origin of the right coronary artery; an anomalous right coronary artery, with an intra-arterial and intramural course may appear to have a normal origin in this view, when the coronary is seen in cross-section coursing within the wall of the aorta. The level of origin of the left coronary artery may be seen in the apical five chamber and subcostal left anterior oblique views. The parasternal long axis view may also be used to assess the left anterior descending artery, as it courses along the anterior interventricular groove. This is best accomplished by tilting the transducer towards the patient's left and anteriorly, as sweeping from the aorta to the main pulmonary artery.

7. Conclusion

Transthoracic echocardiography is the initial imaging modality used to screen for congenital coronary anomalies in the pediatric population. Unfortunately, during many pediatric echocardiograms the coronary arteries are not well seen or incompletely evaluated. This review discusses a standardized approach to evaluating coronary arteries and techniques for image optimization. We hope this may be useful as a concise reference on basic coronary artery imaging. A standardized transthoracic imaging protocol performed by an experienced echocardiographer has been shown to improve detection rates of anomalous aortic origin of the coronary artery from an inappropriate sinus of Valsalva. A thorough evaluation includes determining the coronaries' sinus of origin, the level of origin (“take-off”) from the aorta, their proximal course, visualization of flow within the coronary arteries, and

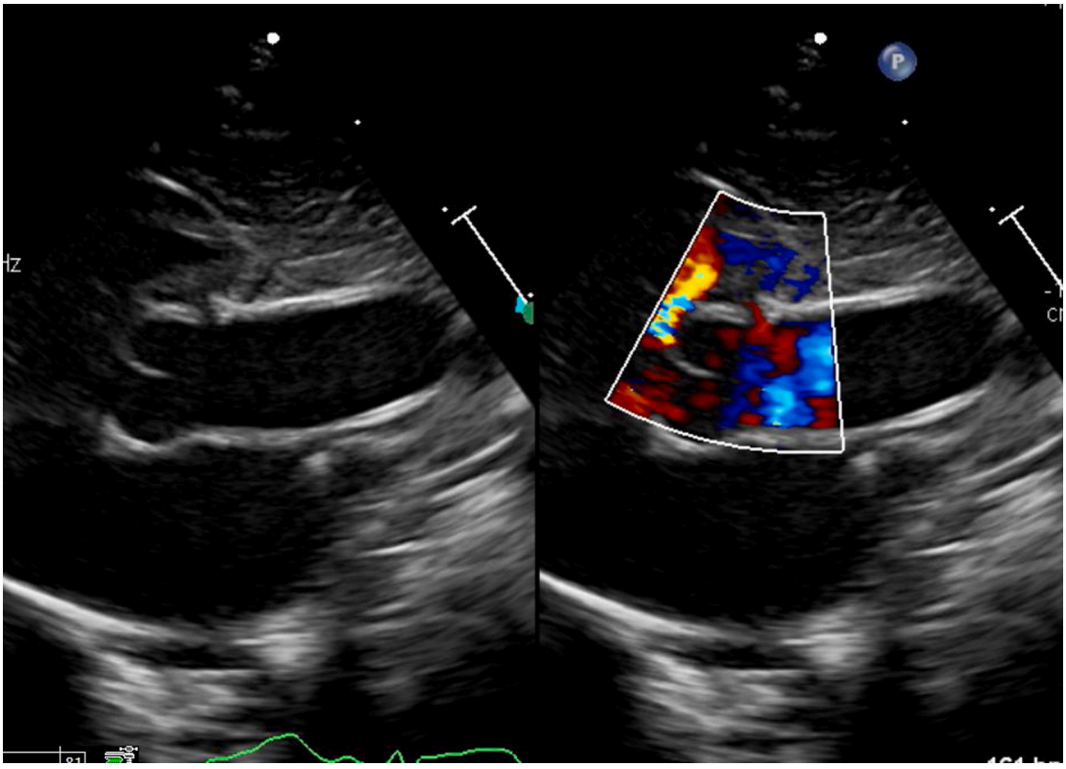

Fig. 4. Normal level of origin of the right coronary artery as seen in the parasternal long axis view.

assessment of their directionality of flow. We recommended the following coronary imaging settings: use the highest frequency probe suitable for each patient, the color scale should be reduced to 20–30 cm/s during color Doppler evaluation, a small pie shaped sector should be used, the frame rate should be above 30 Hz, lower compression setting (35–40) and two dimensional gain setting (50–60%), high persistence, and high flow optimization. In some disease states, measurement of the coronary artery diameter from inner edge to inner edge may also be indicated. Cine clips of two-dimensional and color Doppler imaging should be recorded. Our institution prefers this to be performed using color compare feature of the equipment in addition to separate two dimensional and color images of the coronary arteries. Still frame images of the coronary arteries are not recommended because false dropout may cause an anomalous coronary artery to appear normal. We recommend interrogating the area between aorta and the pulmonary artery in parasternal short-axis view with color Doppler to identify diastolic color flow. Acute angle takeoff in addition to the color flow within proximal coronary lumen can be used to make the diagnosis of a proximal intramural course of anomalous coronary artery [5].

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Credit author statement

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Laura Cavallero: Writing: original draft, reviewing and editing.
Nivedit Kudchadker: Conceptualization, Writing: reviewing and editing, supervision and final submission.

Declaration of competing interest

The authors have no conflict of interest.

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