Appendix 2. Routine Screening Protocols During Pregnancy at Toronto HHT Centre

Patients consulting for the first time at the HHT (Hereditary Hemorrhagic Telangiectasia) Centre during pregnancy, and therefore not previously screened for pulmonary arteriovenous malformations (AVMs), are screened during early second trimester with an arterial blood gas analysis and transthoracic contrast echocardiography. If either test is abnormal, a low dose unenhanced computer tomography (CT) chest is performed, as soon as possible, during second or third trimester. If the CT demonstrates significant pulmonary AVMs, limited pulmonary angiography with embolization of pulmonary AVMs is recommended. Pregnant women with a previously treated pulmonary AVM are followed during pregnancy, with arterial blood gas analysis in second and third trimesters. Pregnant women with diagnosed but untreated small pulmonary AVMs are followed with the same protocol. If worsening shunt is detected by arterial blood gas analysis, low-dose unenhanced CT chest is recommended, as soon as possible, in second or third trimester. If the CT demonstrates significant pulmonary AVMs, limited pulmonary angiography with embolization of pulmonary AVMs is recommended.

When pulmonary angiography and embolization are performed during pregnancy, we use standard technique as previously described^{1,2} but with shielding of the fetus, avoiding fluoroscopy over the groin, and minimizing fluoroscopy time. In these cases we do not perform full bilateral pulmonary angiography but rather focus on known pulmonary AVMs identified from CT.

Cerebral AVM screening is recommended and is performed either in early third trimester (unenhanced MRI) or early postpartum. Spinal AVM screening is generally not performed during pregnancy.

de Gussem EM, Lausman AY, Beder AJ, Edwards CP, Blanker MH, Terbrugge KG, et al. Outcomes of pregnancy in women with hereditary hemorrhagic telangiectasia. Obstet Gynecol 2014;123.

The authors provided this information as a supplement to their article.

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