Title: Early identification and screening guidelines for monochorionic twins with TAPS **Date**: March 24th, 2021

Executive summary:

Over the past thirty years, the incidence of twin births has nearly doubled due to the use of assisted reproductive technology and advanced maternal age. Monochorionic (identical) twins account for about 20% of all twin pregnancies, and compared to dichorionic (fraternal) twins, they are at an increased risk for numerous complications, including preterm birth, growth restriction, and stillbirth. In addition, monochorionic twins can share a placenta, which provides nutrients and oxygen to the baby, however this can also lead to the development of abnormal connections between blood vessels in the placenta. When the connections between the blood vessels are large enough, it leads to a rapidly progressive disorder called twin to twin transfusion syndrome (TTTS). This leads to one twin being supplied more blood, nutrients, and oxygen from the placenta. This disorder is considered a rare disease by the NIH and is commonly monitored in monochorionic pregnancies. However, there is another disorder called twin anemia-polycythemia sequence (TAPS) which also affects monochorionic twins and occurs when abnormal tiny surface connections in the placenta form leading to a slow transfusion of red blood cells from one twin to the other twin. Unlike TTTS, it leads to anemia in one twin and polycythemia in the other twin. Unfortunately, due to the lack of guidelines and awareness for the disorder, there are many missed TAPS diagnoses, and if the disease is not treated, the babies could be born with severe birth defects, intellectual disability, or be unfortunately stillborn. Thus, it is necessary to establish guidelines for diagnosing, screening, and early referral to a maternal-fetal physician to support all families with monochorionic twins.

Scope of problem:

Due to the lack of awareness/recognition of international research and inconsistent guidelines over the past 15 years, TAPS is a relatively undiagnosed disorder. Spontaneous TAPS can occur in 3-5% of monochorionic twins, however it can also occur in 2-16% of twins following laser surgery for the more well-known disorder, TTTS. Importantly, it has not been viewed as a separate disorder from TTTS by the NIH Office of Rare Disease (NORD) and failure to distinguish TAPS independently from TTTS limits physician and patient awareness. Furthermore, the current recommendation from the Society for Maternal Fetal Medicine (from 2013) does not support regular screening for TAPS due to lack of information in fetal outcomes~~,~~ which limits patient access to screening and potential interventions.

Policy alternatives:

- Recognition of TAPS as a rare disorder and a separate disease from TTTS by NORD/ICD-10-CM.
  - ***Advantages:*** This would increase TAPS awareness and potentially provide additional research funding for TAPS, early intervention, and contribute to better fetal outcomes. This would also require the Society for Maternal Fetal Medicine to update their guidelines and standardize the level of care provided to all monochorionic pregnancies.
  - ***Disadvantages:*** There is little information on the pathophysiology of TAPS and TTTS so the conditions may be similar.
- Upon identification of monochorionic twins, immediate referral to a maternal fetal doctor due to high-risk pregnancy.
  - ***Advantages:*** This will provide specialized care for high-risk pregnancies and increased screening for rare and disabling disorders. Moreover, surviving infants of TAPS will also be able to receive specialized follow-up care.
  - ***Disadvantages:*** Increased number of referrals could unnecessarily increase physicians' caseload and overutilize resources.
- Implement monochorionic twin screening guidelines from International Society of Ultrasound in Obstetrics & Gynecology, which is measuring the middle cerebral artery peak systolic velocity via Doppler screening every 2 weeks for all monochorionic twins starting at 16 weeks gestation.
  - ***Advantages:*** The guidelines should standardize screening for all monochorionic twin pregnancies, which can lead to early intervention for TAPS. With early diagnosis and screening, future research can be established to improve fetal outcomes.
  - ***Disadvantages:*** There is still limited knowledge of the utility of increased screenings for TAPS and improved fetal outcomes, thus standardized screening, could lead to overuse of resources without a confirmation of improved outcomes. Additional appointments may be costly for patients as well.

Recommendations:

With the increasing incidence of twin pregnancies and almost 20% of those pregnancies being monochorionic, there is an increased need for improving the standard of care for all monochorionic pregnancies. Thus, immediate referral to a specialized maternal fetal doctor for additional monitoring would improve maternal and fetal outcomes. Current guidelines do not support the screening of TAPS, and TAPS is not listed as a separate disorder from TTTS within NORD/ICD-10-CM. However, additional research over the past 15 years identifies a need for regulation and standardization of screening for TAPS. Thus, updating the guidelines to include TAPS screening and separating TAPS from TTTS will raise awareness in this rare but disabling disorder and will increase funding for research into the causes, diagnosis, and treatment of TAPS.

References:

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