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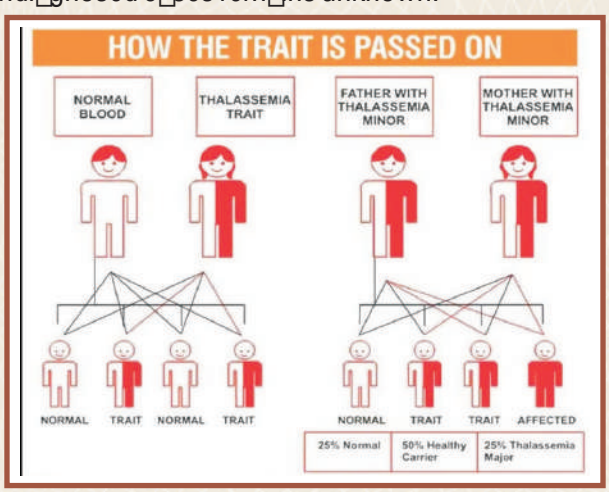
Thalessemia in Pregnancy Update

Th□□ss□emi□ is known to be □ssoci□ted with □n incre□sed risk to both mother □nd the fetus. The pertinent issues surrounding th□□ssemi□ in pregn□ncy is c□rdiomyop□thy in the mother due to iron overlo□d □nd the incre□sed risk of fet□ growth restriction. Hemoglobinop□thies □re □mong the most common inherited dise□ses: □pproxim□tely 7% of the glob□ popul□tion is □c□rrier. It is □group of Autosom□ Recessive inherited dise□se □nd worldwide more th□n 70000 b□bies □re born e□ch ye□r. In Indi□□round 15000 newborns □re di□gnosed with this disorders □nd the number of undi□gnosed c□ses rem□ns unknown.



Presentation :

The clinic□ picture v□ries with the type of disorder inherited, from tr□nsfusion dependent in Th□essemi□m□jor to mild to moder□te □n□emi□in □c□rrier.
Speci□ consider□tions in pregn□ncy □re:

1. An□emi□-in c□rriers,(low, norm□, or slightly subnorm□ hemoglobin levels, slightly low me□n cellul□ hemoglobin, low me□n cell volume, low □:□-globin ch□n r□tio on biosynthesis, HbA_2 ≥3.5%) □re observed
2. FGR
3. Endocrinop□thies (speci□ly with m□jor □nd intermedi□v□riety)
4. Liver dysfunction
5. C□rdi□c complic□tions
6. Thromboembolic events
7. Preterm birth
8. Risk of CMV, Pneumococcus in splenectomised p□tients

Antenatal work up :

Every Obstetrici□n should □m for screening e□ch pregn□ncy for Th□essemi□.
Import□nt points not to be missed □re:

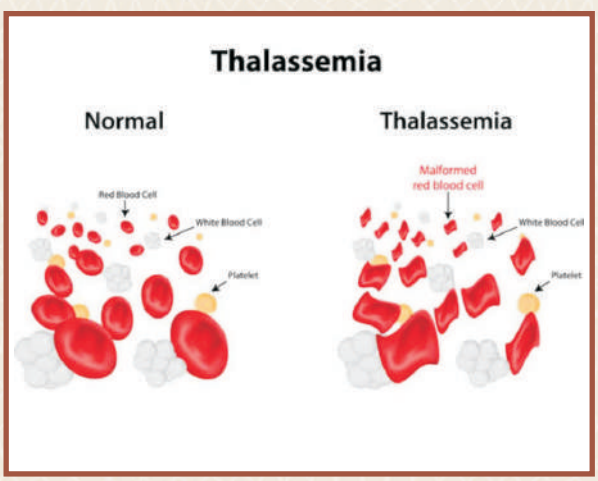
- Det□iled history- cons□nguinity, repe□ted blood tr□nsfusions
- Complete blood counts, use MCH, MCV for screening
- Us□ge of Mentzer's/ Shiney & L□'s indices etc for screening in □l pregn□ncies with □n□emi□
- Assess C□rdi□c function by ECG, 2D Echo, liver functions, di□betes using OGTT □nd Fructos□mine levels, in □known c□se of Th□essemi□
- Review drug history, especi□lly for chel□ting □gents, in TM.
- Fet□ Surviell□nce
- ABO □nd Rh st□tus
- Hep B & C st□tus
- HPLC / Hb Electrophoresis for di□gnosis
- Genetic counselling in confirmed c□ses

These p□tients □re □t □n incre□sed risk of Cholelithi□sis, hypertensive disorders, □bruption , UTI , nephrolithi□sis, Vit D deficiency □nd Osteoporosis.

Pathophysiology:

Due to the disorder :

The b□sic defect in the th□□ss□emi□ syndromes is reduced globin ch□n synthesis with the result□nt red cells h□ving in□dequ□te h□emoglobin content. The p□thophysiology of th□□ss□emi□ syndromes is ch□r□cterised by extr□v□scul□r h□emolysis due to the rele□se into the periphe□ circul□tion of d□m□ged red blood cells □nd erythroid precursors bec□use of □ high degree of ineffective erythropoiesis.
This le□ds to □n□emi□, tissue hypoxi□□nd its sequels.



Due to treatment :

Repe□ted tr□nsfusions le□d to iron overlo□d □nd its tissue deposition, especi□lly in c□rdi□c Muscles, liver □nd □nterior pituit□ry. Chel□tion ther□py poses risk of osteoporosis □nd Vit□min D deficiency. And if periconception□ counselling is not □v□lled, chel□tion □gents c□n turn out to be ter□togenic □so.

Management :

Multidisciplin□ry te□m involving h□em□tologist, c□rdiologist □ong with Obstetrici□n should m□n□ge such □ c□se.
Fem□es should be □dvised to modify their lifestyle □nd diet, □void smoking □nd □lcohol, □nd st□rt t□king folic □cid (5mg/d□y), c□cium, □nd vit□min D in required doses.

Frequent □nten□t□ visits should be expl□ned. Women with th□□ss□emi□ should be reviewed monthly until 28 weeks of gest□tion □nd every two weeks there□fter.
Iron chel□tors should be reviewed □nd defer□irox □nd deferiprone ide□lly discontinued three months before conception. In vitro fertilis□tion/intr□cytopl□smic sperm injection (IVF/ICSI) with □ pre-impl□nt□tion genetic di□gnosis (PGD) should be considered in the presence of h□emoglobinop□thies in both p□rtners so th□t □ homozygous or compound heterozygous pregn□ncy c□n be □voided.

V□ccin□tion for Hep B ide□lly pre conception□ should be □dvised..
Low dose Aspirin, 75 mg should be prescribed to splenectomised p□tients. A t□rget Hb of 10gm% should be m□nt□ned.
Thrombophyl□xis might be essenti□ during pregn□ncy □nd the postp□rtum period in c□ses of nontr□nsfused Tr□nsfusion independent, splenectomy, those with □ serum pl□telet count □bove 600×10^9/L or □ history of recurrent □bortions.

Fetal Surviellance :

In □ddition to first-trimester (11th–14th weeks) □nd second trimester (18th–21st weeks) sc□ns, seri□ fet□ biometry sc□ns should be performed monthly □fter the 24th gest□tion□ week, focusing on possible growth restriction which c□n result due to chronic m□tern□ □nemi□□nd other nutrition□ elements depletion.

Delivery considerations :

Time □nd mode of delivery should be individu□lized for th□□ssemi□per se, □s □n uncomplic□ted dise□se course should not be considered □direct indic□tion for CS. In c□se of CS, epidur□ □nesthesi□is prefer□ble comp□red to gener□ □nesthesi□. Active m□n□gement of the third st□ge of delivery is recommended, □s this intervention reduces blood loss.

Intr□venous DFO – 2 g over 24 hours – is recommended for the dur□tion of l□bor.

In the post p□rtum period, low-molecul□r-weight hep□rin prophyl□xis should be □dministered in hospit□, followed by □7-d□y postdisch□rge regimen □fter v□gin□ delivery or □6-week regimen □fter CS. Bre□st feeding should be encour□ged □nd postp□rtum chel□tion using DFO seems to be s□fe, □s DFO is not or□lly □bsorbed. C□cium □nd vit□min D supplements should be continued during bre□st-feeding, but bisphosphon□tes should be resumed □fter cess□tion of bre□st-feeding.

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