1. Multiple Placental Infarcts in a Pregnant Woman with Essential Thrombocythemia.

**Author(s):** Edahiro, Yoko; Ando, Jun; Suzuki, Toshifumi; Fukumura, Yuki; Masuda, Azuchi; Sakayori, Shiori; Takeda, Jun; Maruyama, Yojiro; Makino, Shintaro; Itakura, Atsuo; Komatsu, Norio

**Source:** Internal medicine (Tokyo, Japan); Dec 2018; vol. 57 (no. 24); p. 3647-3650

**Publication Date:** Dec 2018

**Publication Type(s):** Journal Article

**PubMedID:** 30101937

Available at [Internal medicine (Tokyo, Japan)](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6440799/) - from Europe PubMed Central - Open Access

**Abstract:** Myeloproliferative neoplasms (MPNs), including polycythemia vera, essential thrombocythemia (ET), and primary myelofibrosis, mainly occur in older patients, but have also been reported in younger patients. A "second peak" occurs in female patients in their thirties, particularly in ET; thus, the management of pregnancy is often discussed. We herein present the case of a 33-year-old woman with a high platelet count and multiple placental infarcts during delivery who was subsequently diagnosed with ET. Although there are no worldwide guidelines for the management of MPNs in pregnancy, the risk of thrombosis is markedly increased in these patients, and antithrombotic therapy should be considered.

**Database:** Medline

2. The course of acquired von Willebrand syndrome during pregnancy among patients with essential thrombocytosis.

**Author(s):** Rottenstreich, Amihai; Kleinstern, Geffen; Amsalem, Hagai; Kalish, Yosef

**Source:** Journal of thrombosis and thrombolysis; Oct 2018; vol. 46 (no. 3); p. 304-309

**Publication Date:** Oct 2018

**Publication Type(s):** Journal Article

**PubMedID:** 29654448

Available at [Journal of thrombosis and thrombolysis](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6440799/) - from SpringerLink

**Abstract:** To investigate the course of acquired type 2A von Willebrand syndrome (AVWS) in relation to patient management and outcomes among pregnant patients with essential thrombocytosis (ET). A review of pregnant women with ET evaluated for AVWS at the beginning of pregnancy and at the third trimester. Eighteen women with 24 pregnancies were included in this study. A history of bleeding was noted in 8 (44%) patients. In 20 (83%) pregnancies AVWS was evident at the initial testing. Following initial testing, antithrombotic therapy was administered in 22 (92%) pregnancies (aspirin, n = 20 and low-molecular-weight heparin, n = 2). In the remaining two pregnancies,
VWF:RCo levels were below 30%; thus, aspirin was given only after repeat testing at 14-16 weeks. At third trimester testing, median VWF:RCo levels were significantly higher than at the initial testing (86 vs. 48%, P < 0.001), with no evidence of AVWS in any of the patients. Significant increases were also observed in the VWF:Ag level (127 vs. 84%, P < 0.001), the VWF:RCo/VWF:Ag ratio (0.75 vs. 0.54, P < 0.001) and the FVIII level (103 vs. 68%, P < 0.001); while platelet count (359 vs. 701 × 10⁹/l, P < 0.001) and hemoglobin level (11.6 vs. 13.4 g/dl, P < 0.001) decreased. Neuraxial anesthesia was safely performed in 17 (71%) pregnancies. No significant bleeding events occurred during pregnancy and delivery. AVWS-related abnormalities in women with ET mostly improved during pregnancy, with favorable maternal and fetal outcomes. VWF parameters should be tested at early pregnancy and repeated at the third trimester, to guide pregnancy and delivery management.

Database: Medline

3. Contemporary management of patients with BCR-ABL1-negative myeloproliferative neoplasms during pregnancy.

Author(s): Griesshammer, Martin; Sadjadian, Parvis; Wille, Kai

Source: Expert review of hematology; Sep 2018; vol. 11 (no. 9); p. 697-706

Publication Date: Sep 2018

Publication Type(s): Journal Article

PubMedID: 30084669

Abstract: INTRODUCTION The management of pregnancy during the course of BCR-ABL1-negative myeloproliferative neoplasms (MPN) is an increasingly relevant problem. This is mostly due to earlier and better diagnosis of MPN together with the trend in modern society toward delaying pregnancy until later life. Areas Covered: The present review aims to provide an overview of the available literature data concerning outcome of pregnancy in MPN. Possible therapeutic modalities are discussed and a management algorithm is suggested. Expert Commentary: Most data are available for women with essential thrombocythemia and we present 793 published pregnancies. Live birth rate is 68.5% with 31.5% miscarriages. Spontaneous abortion is the most frequent complication with 26.5%, followed by stillbirth with 4.8%. Maternal complications are relatively low with 1.8% major thrombotic and 2.4% major bleeding events. In polycythemia vera the situation is clinically more complex and roughly 150 pregnancy reports are available. There is very limited information in primary myelofibrosis with less than 20 reported pregnancies. With active management including control of blood counts, aspirin, low molecular weight heparin and in higher risk cases interferon alpha pregnancy in MPN is manageable with a success rate not far below the normal situation with 80%.

Database: Medline
4. Efficacy and safety of interferon alpha for essential thrombocythemia during pregnancy: two cases and a literature review.

**Author(s):** Sakai, Kazuya; Ueda, Akane; Hasegawa, Masaaki; Ueda, Yasunori

**Source:** International journal of hematology; Aug 2018; vol. 108 (no. 2); p. 203-207

**Publication Date:** Aug 2018

**Publication Type(s):** Case Reports Journal Article Review

**PubMedID:** 29290077

Available at [International journal of hematology](https://link.springer.com/article/10.1007/s11238-018-0012-0) - from SpringerLink

**Abstract:** For pregnant women with essential thrombocythemia (ET), no standard approach for managing the platelet count has been established. We present the cases of two pregnant women with ET treated with interferon (IFN)-alpha. Each case showed a marked platelet decrease, from values within normal limits at the time of delivery, with no severe adverse events. To clarify the efficacy and safety of IFN alpha for ET during pregnancy, we performed a literature review. A total of 43 pregnant women with ET were ultimately identified from 12 articles and the present cases. IFN-alpha therapy decreased platelet counts to normal levels at birth in many cases, and there were no adverse events that required the discontinuation of IFN-alpha treatment. Overall, 93% of pregnant women with ET gave birth to healthy babies. We consider that, given its efficacy and safety, IFN-alpha therapy is a reasonable treatment option for pregnant women with ET.

**Database:** Medline

5. Essential thrombocythemia treatment algorithm 2018

**Author(s):** Tefleri A.; Vannucchi A.M.; Barbui T.

**Source:** Blood Cancer Journal; Jan 2018; vol. 8 (no. 1)

**Publication Date:** Jan 2018

**Publication Type(s):** Article

**PubMedID:** 29321520

Available at [Blood Cancer Journal](https://academic.oup.com/bloodcancerjournal) - from ProQuest (Hospital Premium Collection) - NHS Version

Available at [Blood Cancer Journal](https://academic.oup.com/bloodcancerjournal) - from Europe PubMed Central - Open Access

**Abstract:** Current drug therapy for myeloproliferative neoplasms, including essential thrombocythemia (ET) and polycythemia vera (PV), is neither curative nor has it been shown to prolong survival. Fortunately, prognosis in ET and PV is relatively good, with median survivals in younger patients estimated at 33 and 24 years, respectively. Therefore, when it comes to treatment in ET or PV, less is more and one should avoid exposing patients to new drugs that have not been shown to be disease-modifying, and whose long-term consequences are suspect (e.g., ruxolitinib). Furthermore, the main indication for treatment in ET and PV is to prevent thrombosis and, in that regard, none of the newer drugs have been shown to be superior to the time-tested older drugs (e.g., hydroxyurea). We currently consider three major risk factors for thrombosis (history of thrombosis, JAK2/MPL mutations, and advanced age), in order to group ET patients into four risk categories: "very low risk" (absence of all three risk factors); "low risk" (presence of JAK2/MPL mutations); "intermediate-risk" (presence of advanced age); and "high-risk" (presence of thrombosis history or presence of both JAK2/MPL mutations and advanced age). Herein, we provide a point-of-care treatment algorithm that is risk-adapted and based on evidence and decades of experience. Copyright © 2018 The Author(s).

**Database:** EMBASE

**Author(s):** Puyade, Mathieu; Cayssials, Emilie; Pierre, Fabrice; Pourrat, Olivier

**Source:** Obstetric medicine; Dec 2017; vol. 10 (no. 4); p. 165-169

**Publication Date:** Dec 2017

**Publication Type(s):** Journal Article

**PubMedID:** 29225675

**Available at:** Obstetric medicine - from Europe PubMed Central - Open Access

**Abstract:**
Background: The most frequent myeloproliferative neoplasms are essential thrombocythemia and chronic myelogenous leukemia, which usually manifests with thrombocytosis. Only essential thrombocythemia is associated with morbidity during pregnancy (recurrent miscarriages, intrauterine fetal death, small for gestational age and preeclampsia). The aim of this paper is to describe outcomes of pregnancy in women with myeloproliferative neoplasms seen at a single academic institution.

Methods: Data were collected retrospectively from 2002 to 2015. Descriptive analyses were performed.

Results: Eighteen pregnancies in 13 patients and 17 births were identified. One patient had recurrent miscarriages. There were two intrauterine fetal deaths, three small for gestational age linked to vascular placenta pathology and one preeclampsia. All of these mothers harbored JAK2V617F mutation. Two out of three patients with small for gestational age developed a venous thrombosis in the two years following delivery.

Conclusion: Thrombocytosis associated with myeloproliferative neoplasms should be considered as a risk factor for maternal and fetal complications.

**Database:** Medline

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7. Primary myelofibrosis and pregnancy outcomes after low molecular-weight heparin administration: A case report and literature review

**Author(s):** Elena Bohiltea R.; Mihaela Cirstoiu M.; Turcan N.; Niculescu-Mizil E.; Vladareanu A.M.; Voican I.; Antoniu Ionescu C.; Dimitriu M.

**Source:** Medicine (United States); Nov 2017; vol. 96 (no. 46)

**Publication Date:** Nov 2017

**Publication Type(s):** Review

**PubMedID:** 29145319

**Available at:** Medicine - from Europe PubMed Central - Open Access

Available at Medicine - from Ovid (Journals @ Ovid) - Remote Access

**Abstract:**
Rationale: Primary myelofibrosis is encountered with the myeloproliferative diseases and is the least prevalent among women of childbearing age. The prognosis is guided by pancytopenia, leukemic transformation and thrombosis which are the dominant complications. Patient concerns: Data regarding protocol management during pregnancy in the context of myelofibrosis are insufficient. Fewer than ten cases have been described until now and half of this cases have resulted in fetal death due to placental infarction during the second and third trimesters. Diagnoses: We present the case of a 34-year-old pregnant woman diagnosed with Jak 2- negative primary myelofibrosis. Personal history did not include miscarriage or stillbirth. Interventions: The patient was previously treated with anagrelide hydrochloride, which was interrupted at 6 weeks of gestation when the pregnancy was confirmed. It was replaced with Interferon-a 3 MU/day. Because of severe thrombocytosis, administration of aspirin 150 mg/day was recommended. Outcomes: The pregnancy was uneventful. The patient was hospitalized at 33 weeks of gestation because of moderate vaginal bleeding and high risk of preterm birth. After a specialized hematological investigation, the treatment with aspirin was replaced with low-molecular-weight heparin 0.6 ml per day. This
combined treatment assisted in the natural tendency to lower platelet counts during pregnancy and resulted in stabilization of the hematological status. At 38 weeks of gestation the patient delivered a healthy baby boy via cesarean. He weight 2850 grams and his Apgar score was 9. Anticoagulant and interferon treatments were continued post-partum under hematologist surveillance. Lessons: This case was rare and complex. Because it was related to pregnancy it required continuous collaboration and supervision between obstetrician and hematologist.

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Database: EMBASE


Author(s): Kleman, Ariel; Singavi, Arun K; Michaelis, Laura C

Source: Clinical advances in hematology & oncology : H&O; Oct 2017; vol. 15 (no. 10); p. 773-783

Publication Date: Oct 2017

Publication Type(s): Journal Article Review

PubMedID: 29040257

Abstract: Essential thrombocythemia (ET), an uncommon blood cancer, is one of the classic myeloproliferative neoplasms, a category that also includes polycythemia vera and primary myelofibrosis. All 3 diseases are clonal hematopoietic stem cell disorders. Since 2005, when scientists discovered a molecular aberration driving clonal hematopoiesis in polycythemia vera, our understanding of the genomic underpinnings of these conditions has increased rapidly. Over the last decades, primary prevention of thrombotic and hemorrhagic complications has improved the lives of patients with ET, and the ability to characterize the disease by the presence or absence of molecular mutations has lent precision to our prognostic models. This review outlines a modern approach to the diagnosis and treatment of ET. It highlights the 2016 World Health Organization standards for differentiating the disease from primary myelofibrosis, which is key for an accurate prognosis. It also describes the current risk stratification models and discusses the vascular and hemorrhagic risks that affect patients with this chronic condition, including younger individuals and pregnant women. Finally, it outlines a simple-to-follow treatment algorithm that is based on an understanding of the vascular risks and provides a foundation for discussing treatment choices with patients.

Database: Medline

**Author(s):** Kempen, Paul Martin

**Source:** A & A case reports; Sep 2017; vol. 9 (no. 6); p. 172-174

**Publication Date:** Sep 2017

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 28520566

Abstract: Essential thrombocytosis (ET) is a rare disease with known thrombotic and bleeding complications. We encountered a patient with a diagnosis of longstanding Janus kinase-2 gene-negative ET on aspirin therapy presenting for labor epidural. Evaluation of platelet function with point-of-care analysis using Plateletworks in a community hospital setting allowed confirmation of adequate numbers of functional platelets to support safe epidural placement. The relevant issues of ET for anesthesia management with labor epidurals are discussed. Unique, relevant, and unexpected findings from the platelet function testing are presented.

**Database:** Medline

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10. Management of MPN in pregnancy

**Author(s):** Griesshammer M.

**Source:** Oncology Research and Treatment; Sep 2017; vol. 40; p. 33-34

**Publication Date:** Sep 2017

**Publication Type(s):** Conference Abstract

Abstract: Introduction: Myeloproliferative neoplasms (MPN), essential thrombocytopenia (ET), polycythemia vera (PV) and primary myelofibrosis (PMF) often present in women of childbearing age. The course of MPN is associated with an increased incidence of microcirculatory, hemorrhagic and thromboembolic complications. As both MPN and pregnancy are associated with a higher incidence of thromboembolic complications, a dramatic increase in such complications is a matter concern. If a MPN woman gets pregnant these complications may affect both the mother and the fetus. Methods: In the literature, most pregnancies are reported for women with ET and there are fewer reports in women with polycythemia vera PV. As information about pregnancies in myelofibrosis is very limited this paper is focusing on pregnancies in ET and PV. Results: In our literature review we report on a total of 730 pregnancies in 463 women with ET. The success rate (baby alive) is 69%, the rate of miscarriage is 31%. First trimester abortion was the most frequent complication and occurred in 26%. A full-term normal delivery, including forceps delivery or caesarean section, was seen in 61%. The cumulative incidence of major maternal complications was 1.3% for major thromboembolic events and 3% for major bleeding. Within the European LeukemiaNet we collected 121 preg in 48 PV patients. The success rate of pregnancies was significantly better (49% versus 77%, respectively) for patients in whom the diagnosis of PV was known and appropriate management performed according to current guidelines. In high risk ET and PV patients all patients should be treated with lownoise aspirin (50 to 100mg/day) plus interferon alpha throughout pregnancy. The immediate puerperium is indeed the time of greatest risk for venous thrombosis for which prophylaxis with low molecular weight heparin is indicated and should be given for at least six weeks post-partum. Conclusion: With an appropriate risk stratified management encouraging data are available regarding pregnancy outcomes in both ET and PV patients and a medical abortion is rarely indicated.
11. Risk of venous thromboembolism in pregnant women with essential thrombocythemia: a systematic review and meta-analysis.

Author(s): Skeith, Leslie; Carrier, Marc; Robinson, Susan E; Alimam, Samah; Rodger, Marc A

Source: Blood; Feb 2017; vol. 129 (no. 8); p. 934-939

Publication Date: Feb 2017

Publication Type(s): Meta-analysis Journal Article Review Systematic Review

PubMedID: 28049641

Abstract: We performed a meta-analysis to evaluate the risk of venous thromboembolism (VTE) in pregnant women with essential thrombocythemia. Twenty-one trials and 756 pregnancies met inclusion criteria. The absolute VTE risk in the antepartum period is not above a threshold where low-molecular-weight heparin (LMWH) prophylaxis is clearly indicated or below a threshold where LMWH should be withheld (2.5%; 95% CI, 1.3-4.3). Postpartum, the absolute VTE risk is above a threshold where postpartum LMWH prophylaxis should be considered (4.4%; 95% CI, 1.2-9.5).

Database: Medline

12. A Successful Mother and Neonate Outcome for a Woman with Essential Thrombocytosis and FV Leiden Heterozygosity.

Author(s): Politou, Marianna; Valsami, Serena; Gkorezi-Ntavela, Irontianta; Telenis, Vasilios; Merkouri, Efrosyni; Christopoulos, Panagiotis

Source: Case reports in obstetrics and gynecology; 2016; vol. 2016; p. 7041686

Publication Date: 2016

Publication Type(s): Journal Article

PubMedID: 27123352

Abstract: Essential thrombocytosis (ET) and FV Leiden heterozygosity represent an acquired and hereditable hypercoagulable state, respectively. An uncommon case of coexistence of ET and FV Leiden heterozygosity in a 36-year-old pregnant woman and her successful pregnancy outcome is described. She was considered to be at high risk of thrombosis during her pregnancy and she was treated with both prophylactic dose of LMWH and aspirin daily throughout her pregnancy and for a 6-week period postpartum. The efficacy of the anticoagulation treatment was monitored in various time points not only by measuring anti-Xa levels and D-Dimers but also with new coagulation methods such as rotation thromboelastometry and multiplate. Global assessment of coagulation using additional newer laboratory tests might prove useful in monitoring coagulation pregnancies at high risk for thrombosis.

Database: Medline

**Author(s):** Alimam, Samah; Bewley, Susan; Chappell, Lucy C; Knight, Marian; Seed, Paul; Gray, Gabriella; Harrison, Claire; Robinson, Susan

**Source:** British journal of haematology; Oct 2016; vol. 175 (no. 1); p. 31-36

**Publication Date:** Oct 2016

**Publication Type(s):** Multicenter Study Journal Article

**PubMedID:** 27612319

**Abstract:** The reported higher risk of maternal and fetal complications in women with myeloproliferative neoplasms (MPN) poses challenge during pregnancy. A national prospective study of maternal and fetal outcomes of pregnant women with a diagnosis of MPN was undertaken via the United Kingdom Obstetric Surveillance System between January 2010 and December 2012. Fifty-eight women with a diagnosis of MPN were identified; 47 (81%) essential thrombocythaemia, five (9%) polycythaemia vera, five (9%) myelofibrosis and one (2%) MPN-unclassified. There were 58 live births. The incidence of miscarriage was 1.7/100 (95% confidence interval [CI]: 0.04-9.24) and the perinatal mortality rate was 17/1000 (95% CI: 0.44-92.36) live and stillbirths. Incidence of maternal complications was 9% (5/57) pre-eclampsia, 9% (5/57) post-partum haemorrhage and 3.5% (2/57) post-partum haematoma. There were no maternal deaths or thrombotic events. Delivery was induced in 45% (24/53) of women and the Caesarean section rate was 45% (24/53). The majority (85%, 45/53) delivered at term (>37 weeks gestation). Twenty-two percent (12/54) of neonates were below the 10% centile for growth and 13% (7/54) required admission to a neonatal care-unit; there were no neonatal deaths. The findings of this large, UK prospective study suggests women with MPN appear to have successful pregnancies with better outcomes than would be anticipated from the literature.

**Database:** Medline

14. Haematological malignancies in pregnancy: An overview with an emphasis on thrombotic risks

**Author(s):** Horowitz N.A.; Lavi N.; Nadir Y.; Brenner B.

**Source:** Thrombosis and Haemostasis; Oct 2016; vol. 116 (no. 4); p. 613-617

**Publication Date:** Oct 2016

**Publication Type(s):** Article

**PubMedID:** 27465578

**Abstract:** With increase of maternal age, the incidence of haematological malignancies during pregnancy is rising and posing diagnostic and treatment challenges. Lymphoma is the fourth most common malignancy diagnosed in pregnancy; Hodgkin lymphoma is more frequent in pregnant women than non-Hodgkin lymphoma (NHL). The proportion of highly aggressive lymphomas in pregnant women is significantly higher than in non-pregnant women of reproductive age. Reproductive organ involvement is observed in almost half of pregnant women with NHL. The association of acute leukaemia and pregnancy is infrequent and it is assumed that pregnancy does not accelerate the disease course. Both cancer and pregnancy induce a procoagulant state which can lead to maternal venous thromboembolism (VTE) and placental occlusion. Pregnancy in woman with myeloproliferative neoplasms (MPN) promotes thrombotic environment, associating with an augmented risk of placental thrombosis, intrauterine growth retardation or loss and maternal thrombotic events. Haematological malignancies during pregnancy often require urgent diagnosis and management and are associated with potential adverse fetal outcomes. Most chemotherapeutic
agents are teratogenic and should be avoided during the first trimester. Their use during the second and third trimesters may cause intrauterine growth restriction, premature birth and intrauterine fetal death. All chemotherapeutic drugs should be administered only after a detailed discussion with the patient and with close fetal monitoring. Chemotherapy and biological agents might also augment thrombotic risk. Guidelines for VTE prophylaxis in pregnant women with hematologic malignancies, apart from MPN, are currently unavailable, and therefore, clinical judgment should be made in each case. Copyright © Schattauer 2016.

**Database:** EMBASE

15. Myeloproliferative neoplasms in pregnancy: ways to go
**Author(s):** Lavi N.; Brenner B.  
**Source:** British Journal of Haematology; Oct 2016; vol. 175 (no. 1); p. 7-9  
**Publication Date:** Oct 2016  
**Publication Type(s):** Editorial  
**PubMedID:** 27601254  
Available at [British Journal of Haematology](https://onlinelibrary.wiley.com/doi/10.1111/bjh.14611) - from Wiley Online Library Science, Technology and Medicine Collection 2017  
**Database:** EMBASE

**Author(s):** Villani, Michela; Colaizzo, Donatella; Tiscia, Giovanni L; Chinni, Elena; Bodenizza, Carl'Antonio; Cascavilla, Nicola; Grandone, Elvira  
**Source:** Blood coagulation & fibrinolysis: an international journal in haemostasis and thrombosis; Sep 2016; vol. 27 (no. 6); p. 727-728  
**Publication Date:** Sep 2016  
**Publication Type(s):** Case Reports Journal Article  
**PubMedID:** 26650457  
Available at [Blood coagulation & fibrinolysis: an international journal in haemostasis and thrombosis](https://www.sciencedirect.com/science/journal/09576501) - from Ovid (LWW Total Access Collection 2015 - Q1 with Neurology)  
**Abstract:** Coagulation disorder associated with essential thrombocythemia may exacerbate the prothrombotic state physiologically occurring during pregnancy. We report a case of a severe postpartum haemorrhage in a 35-year-old woman previously diagnosed with essential thrombocythemia and carrying the somatic calreticulin mutation. She was referred to our Thrombosis and Haemostasis Unit for pregnancy management. A treatment with low-dose aspirin was prescribed until the labour started, as the platelets count raised above 1000 × 10^3/l. At the time of bleeding, no residual placenta was detected at the revision of the uterine cavity. Although the postpartum is a high-risk period for thrombotic events, we have to carefully evaluate in women with essential thrombocythemia the likelihood of developing a hemorrhagic complication.  
**Database:** Medline
17. Manage haematological malignancies during pregnancy according to cancer type and gestational stage

**Author(s):** Adis Medical Writers  
**Source:** Drugs and Therapy Perspectives; Jul 2016; vol. 32 (no. 7); p. 287-291  
**Publication Date:** Jul 2016  
**Publication Type(s):** Article  
Available at Drugs and Therapy Perspectives - from ProQuest (Hospital Premium Collection) - NHS Version  
**Abstract:** Haematological malignancies that occur during pregnancy are difficult to diagnose and manage. For patients diagnosed during trimester 1, watchful waiting or bridging therapy are options for some types of malignancies, whereas termination should be considered in patients with highly aggressive cancers that require immediate treatment with highly teratogenic agents. The use of chemotherapy is generally considered safe during trimesters 2 and 3. Special considerations should also be given to appropriate supportive care and the timing of delivery. Copyright © 2016, Springer International Publishing Switzerland.  
**Database:** EMBASE


**Author(s):** Beauverd, Yan; Radia, Deepti; Cargo, Catherine; Knapper, Steve; Drummond, Mark; Pillai, Arvind; Harrison, Claire; Robinson, Susan  
**Source:** Haematologica; May 2016; vol. 101 (no. 5); p. e182  
**Publication Date:** May 2016  
**Publication Type(s):** Letter Case Reports  
**PubMedID:** 26819057  
Available at Haematologica - from Europe PubMed Central - Open Access  
**Database:** Medline
19. Risk of pregnancy complications and effect of different treatments in women with essential thrombocythemia: A retrospective monocenter analysis of 62 pregnancies

**Author(s):** Betti S.; Rossi E.; Za T.; Ciminello A.; Bartolomei F.; Chiusolo P.; De Stefano V.

**Source:** Haematologica; Jun 2015; vol. 100 ; p. 534-535

**Publication Date:** Jun 2015

**Publication Type(s):** Conference Abstract

**Abstract:** Background: A high rate of obstetric complications occurs in women with essential thrombocythemia (ET), with a consistent high risk of early fetal loss and a probability of live births around 60%. A variety of therapeutic strategies has been proposed, with a risk-driven intensity of treatment. The role of the JAK2 V617 mutation as predictor of pregnancy complications and possible therapeutic driver is controversial. Aims: To assess in a retrospective cohort of ET patients the JAK2 V617F-related risk of pregnancy complications and the efficacy of different treatments during antepartum and postpartum periods. Methods: We analysed 62 pregnancies occurred in 38 women with ET (median 2 pregnancies per woman, range 1-3) during the years 2001-2014. The median age of the patients at diagnosis was 29 years (range 18-41); the age at conception was >35 years in 33 pregnancies (53%). Nineteen women (50%) carried the JAK2V617F mutation. Two of them had hepatic vein thrombosis and TIA before first conception, respectively. One terminated pregnancy, one blighted ovum and one miscarriage due to Turner syndrome were excluded from further analysis. Antepartum treatment consisted of low molecular weight (LMWH)+aspirin (ASA) in 32 pregnancies, ASA in 14, LMWH in 6; seven pregnancies were left untreated. Interferon was administered during 9 pregnancies. The impact of different antepartum therapeutic strategies was estimated by a multivariate proportional hazards regression model over the weeks of gestation. Puerperium periods were defined as 6 weeks after delivery at >=20 week of gestation and were treated with LMWH in 45 of 51 cases. Results: Among the evaluable pregnancies the rate of live births was 83% (49/59); miscarriage occurred in 8, stillbirth in 2, abruptio placentae with neonatal death in 1, intrauterine fetal growth retardation in 6. Overall, 17 obstetric complications (OC) were considered ET-related (17/59, 29%) and occurred in 7 cases during LMWH+ASA (22%), in 2 during ASA (14%), 3 during LMWH (50%), and 5 in the untreated pregnancies (71%). No thrombosis complicated antepartum-periods. Antithrombotic treatment reduced by 88% the risk of OC in comparison versus untreated pregnancies (odds ratio, OR 0.12, 95%CI 0.02-0.69). The rate of OC was 43% in pregnancies of JAK2 V617F-positive women (13/30) and 14% in the pregnancies of the JAK2 V617F-negative women (4/29) (OR 4.77, 95%CI 1.33-17.18); however after exclusion of the untreated pregnancies the risk associated with the JAK2 V617F mutation was no more significant (OR 3.33, 95%CI 0.85-13.00). A multivariate proportional hazards regression model including age >35 yrs, JAK2 V617F mutation, and antepartum ASA, LMWH, and interferon, retained only ASA as a variable associated with the outcome (OR for complications 0.28, 95%CI 0.10-0.80, p=0.01). Among the 6 untreated puerperium periods, one was complicated by cerebral vein thrombosis (17%), whereas no thrombosis occurred during the remaining puerperium periods treated with LMWH. Summary and Conclusions: In ET patients JAK2 V617F is associated with an increased risk of OC, which was prevented by treatment; namely, in this cohort antepartum ASA was highly effective in preventing ET-related OC; therefore, LMWH should be reserved only to women with additional risk factors for venous thromboembolism independent of ET. The rate of puerperium-related venous thrombosis is high and prompts LMWH prophylaxis.

**Database:** EMBASE
20. Pregnancy complications predict thrombotic events in young women with essential thrombocytopenia.

Author(s): Randi, Maria Luigia; Bertozzi, Irene; Rumi, Elisa; Elena, Chiara; Finazzi, Guido; Vianelli, Nicola; Polverelli, Nicola; Ruggeri, Marco; Vannucchi, Alessandro Maria; Antonioli, Elisabetta; Lussana, Federico; Tieghi, Alessia; Iurlo, Alessandra; Elli, Elena; Ruella, Marco; Fabris, Fabrizio; Cazzola, Mario; Barbui, Tiziano

Source: American journal of hematology; Mar 2014; vol. 89 (no. 3); p. 306-309

Publication Date: Mar 2014

Publication Type(s): Research Support, Non-u.s. Gov't Multicenter Study Journal Article

PubMedID: 24265194

Available at American journal of hematology - from Wiley Online Library Science, Technology and Medicine Collection 2017

Available at American journal of hematology - from Unpaywall

Abstract: Although Philadelphia-negative myeloproliferative neoplasms (MPNs) occur typically in middle to advanced age, any age group may be affected, posing a challenge for their management during pregnancy when they occur in young females. There is a high incidence of thromboembolic events and pregnancy complications in patients with myeloproliferative neoplasms, and a possible relationship between these complications is a matter of concern. The aim of this article was to correlate thrombosis and pregnancy outcome in 158 females with ET experiencing 237 pregnancies. Seven patients had a thrombotic event before their first pregnancy, one of them ended (14.3%) in a miscarriage. Among the 151 patients with no history of thrombosis before they became pregnant, 40 (26.5%) had a miscarriage (P = NS). Eighteen patients (11.4%) developed major thrombotic complications (12 splanchnic vein, 1 cerebral vein, 2 coronary syndromes, and 3 strokes) after at least one pregnancy (4 uneventful and 14 complicated). The occurrence of thrombosis was significantly more frequent (P < 0.001) in patients with a history of pregnancy complications (28%) than in those experiencing a normal pregnancy and delivery (3.7%). Pregnancy complications in women with ET are associated with a higher risk of subsequent thromboses, so pregnant women with this neoplasm who miscarry need to be carefully monitored.

Database: Medline

**Author(s):** Umazume, Takeshi; Yamada, Takahiro; Akaishi, Rina; Araki, Naoto; Nishida, Ryutaro; Morikawa, Mamoru; Minakami, Hisanori

**Source:** Thrombosis research; Feb 2014; vol. 133 (no. 2); p. 158-161

**Publication Date:** Feb 2014

**Publication Type(s):** Journal Article

**PubMedID:** 24268792

Available at [Thrombosis research](https://www.unpaywall.org) - from Unpaywall

**Abstract:**

**INTRODUCTION**
The risk of abortion is known to be high in women with essential thrombocythemia (ET). However, a few studies have focused on the risk of stillbirth among fetuses reaching gestational age compatible with life.

**METHODS**
Review of medical charts of pregnant women with ET who received cares at a single center between January 2003 and June 2013 and the English literature in which more than 20 pregnancies with ET were dealt with regarding outcomes. Outcomes were classified into three categories: spontaneous abortion or preterm delivery before GW 24, stillbirth at and after GW 24, and live birth (LB). Japan national statistics was used to estimate the risk of stillbirth among women with GW ≥22.

**RESULTS**
In all nine pregnancies in four women with ET at our hospital, two miscarriages, one stillbirth (intrauterine death at GW 35), and six LBs occurred. There were six reports in the English literature in which a total of 374 pregnancy outcomes were described: 110 miscarriages (29%), 14 stillbirths (3.7% of all 374 pregnancies and 5.3% of 264 pregnancies with GW≥24), and 250 LBs (67%) occurred. Japan national statistics between 1995 and 2011 indicated that the risk of stillbirth was less than 0.50% among women with GW≥22.

**CONCLUSION**
The risk of stillbirth was extremely high among women with ET. More intensified monitoring of fetal wellbeing may be required to improve outcome of pregnancy complicated with ET.

**Database:** Medline

Author(s): Koh, Mickey B C; Lao, Zhen Tang; Rhodes, Elizabeth

Source: Best practice & research. Clinical obstetrics & gynaecology; Dec 2013; vol. 27 (no. 6); p. 855-865

Publication Date: Dec 2013

Publication Type(s): Journal Article Review

PubMedID: 24060203

Abstract: The management of patients with pre-existing haematological diseases during pregnancy can be particularly challenging. The potential maternal and foetal toxicities from treatment regimens including chemotherapy for malignant haematological disorders mean that joint management between obstetricians and haematologists is essential for achieving good outcomes for both mother and baby. Patients with inherited or acquired disorders of haemostasis including platelets (essential thrombocythaemia) and coagulation (antiphospholipid syndrome) resulting in a prothrombotic state also require special consideration as pregnancy is generally considered to be a prothrombotic condition which could exacerbate the pre-existing disorder. The choice, timing and duration of anticoagulation or anti-platelet therapy require careful coordination during the antenatal, perinatal as well as postnatal periods to ensure that both maternal and foetal risks are taken into consideration. Pregnancy in women with sickle cell disease has long been identified as high risk with medical and pregnancy related risks being more common compared to women without it. A range of foetal risks have also been reported but improvement in outcomes has been seen with better obstetric and haematological care and the emphasis on multidisciplinary teamwork. The meticulous management of iron overload and risks associated with repeated blood transfusions extends into the care of pregnant women with other haemoglobinopathies like thalassemias.

Database: Medline

23. Management of pregnant women with myeloproliferative neoplasms

Author(s): Lavi N.; Avivi I.; Brenner B.

Source: Thrombosis Research; Jan 2013; vol. 131

Publication Date: Jan 2013

Publication Type(s): Article

PubMedID: 23452731

Abstract: Myeloproliferative neoplasms (MPNs) are generally considered to be diseases of elderly population; however, 20% of subjects diagnosed with ET are younger than 40 years. Increase in gestational age in the Western world and improved diagnostic tools raise MPN incidence during pregnancy. MPNs are associated with a remarkable risk for thrombosis and the hypercoagulability milieu associated with pregnancy increases that risk even further. Pregnancies of women diagnosed with MPNs may be complicated with placental thrombosis, fetal growth restriction or loss, and increased risk for maternal thrombosis. The live birth rate in ET and PV is as low as 60%, with first-trimester loss occurring in 20-30% of pregnancies and an increase in late placenta-mediated complications. Major maternal complications (thromboembolic events and bleeding) are more frequent in PV compared with ET. Therapeutic options range from no therapy, aspirin alone, low-molecular weight heparin (LMWH) to cytoreductive therapy, tailored according to patient-specific risk factors. © 2013 Elsevier Ltd. All rights reserved.

Database: EMBASE

Author(s): Singh, Neeta; Kumar, Sunesh; Roy, K K; Sharma, Vaishali; Jalak, Ashish

Source: Platelets; 2012; vol. 23 (no. 4); p. 319-321

Publication Date: 2012

Publication Type(s): Letter Case Reports

PubMedID: 21913812

Available at Platelets - from Unpaywall

Database: Medline


Author(s): Leader, Avi; Pereg, David; Lishner, Michael

Source: Annals of medicine; Dec 2012; vol. 44 (no. 8); p. 805-816

Publication Date: Dec 2012

Publication Type(s): Journal Article Review

PubMedID: 22413913

Abstract: Platelet size correlates with platelet activity and can be assessed by platelet volume indices (PVI). The PVI, mean platelet volume (MPV), is universally available with routine blood counts by automated hemograms and therefore is an attractive index to study in clinical scenarios. PVI are useful in assessing the etiology of thrombocytopenia. In addition, a normal platelet distribution width in the setting of thrombocytosis is highly suggestive of a reactive etiology. Higher MPV is also associated with the presence of cardiovascular risk factors, chest pain due to acute coronary syndrome, and adverse outcome after acute coronary syndrome. Results from studies evaluating MPV in patients with peripheral artery disease, unprovoked deep vein thrombosis, and pulmonary embolism further advocate a potential role for MPV in identifying patients at high risk of thrombosis. Nevertheless, most of these data come from retrospective studies some of which have small study populations and confounding factors influencing platelet volume. Moreover, the cut-off values derived from these retrospective studies have not been validated prospectively. Despite the potential for clinical utility evident from these studies, the above-mentioned flaws together with technical problems in measuring MPV currently limit its clinical usefulness. Our review provides a perspective on PVI's potential clinical use.

Database: Medline
26. Guideline for the diagnosis and management of myelofibrosis

**Author(s):** Reilly J.T.; Mcmullin M.F.; Beer P.A.; Butt N.; Conneally E.; Duncombe A.; Green A.R.; Michael N.G.; Wilkins B.; Harrison C.N.; Gilleece M.H.; Hall G.W.; Mead A.; Knapper S.; Mesa R.A.; Sekhar M.

**Source:** British Journal of Haematology; Aug 2012; vol. 158 (no. 4); p. 453-471

**Publication Date:** Aug 2012

**Publication Type(s):** Article

**PubMedID:** 22651893

Available at British Journal of Haematology - from Wiley Online Library Science, Technology and Medicine Collection 2017

Available at British Journal of Haematology - from Unpaywall

**Abstract:** The guideline group regarding the diagnosis and management of myelofibrosis was selected to be representative of UK-based medical experts, together with a contribution from a single expert from the USA. MEDLINE and EMBASE were searched systematically for publications in English from 1966 until August 2011 using a variety of keywords. The writing group produced the draft guideline, which was subsequently revised by consensus of the members of the General Haematology and Haematology Task Forces of the British Committee for Standards in Haematology (BCSH). The guideline was then reviewed by a sounding board of UK haematologists, the BCSH and the British Society for Haematology Committee and comments incorporated where appropriate. The criteria used to state levels and grades of evidence are as outlined in the Procedure for Guidelines commissioned by the BCSH; the 'GRADE' system was used to score strength and quality of evidence. The objective of this guideline is to provide healthcare professionals with clear guidance on the investigation and management of primary myelofibrosis, as well as post-polycythaemic myelofibrosis (post-PV MF) and post-thrombocythemic myelofibrosis (post-ET MF) in both adult and paediatric patients. © 2012 Blackwell Publishing Ltd.

**Database:** EMBASE
27. A systematic review of the fetal safety of interferon alpha.

**Author(s):** Yazdani Brojeni, P; Matok, I; Garcia Bournissen, F; Koren, G

**Source:** Reproductive toxicology (Elmsford, N.Y.); Jun 2012; vol. 33 (no. 3); p. 265-268

**Publication Date:** Jun 2012

**Publication Type(s):** Journal Article Review Systematic Review

**PubMedID:** 22200624

**Abstract:** BACKGROUND Interferon alpha (IFN) is an effective treatment for a variety of conditions including essential thrombocythemia (ET), chronic myelocytic leukemia, Hepatitis B and C. Because these conditions also occur in women of childbearing age who may become pregnant, information regarding the safety of this medication in pregnancy is essential. This systematic review attempts to summarize all published data on outcome of pregnancies exposed to IFN alpha, trying to differentiate between disease effect and drug effect.

**METHODS** Reports on the use of IFN alpha in human pregnancy and reports on essential thrombocythemia (ET) without use of any medication in pregnancy were identified by a systematic search of the medical literature. We were able to locate only case reports of IFN alpha exposure in pregnancy, of whom 40 out of 63 were diagnosed with ET. We also collected randomly 71 cases (more cases were available in the literature) that were diagnosed with ET due to different etiologies, but who had not received any medication in pregnancy.

**RESULTS** Among the 63 IFN alpha exposures in pregnancy, the mean maternal age was 30±6 years and the mean full term babies' weight was 3096±463 g. Mean gestational age at delivery was 37±3 weeks. There were 55 single and 4 twin pregnancies. No cases of major malformations or stillbirths were reported. There was one case of spontaneous abortion and 13 preterm deliveries (20% of all exposed cases). Among the 71 cases with untreated ET in pregnancy of different etiologies, 46 (65%) had early (within the first 12 weeks of pregnancy) or late (13-20 weeks of gestation) pregnancy loss. There were also 3 cases (4%) of stillbirth and 4 cases (5.6%) of preterm delivery. Only 18 women (25%) delivered healthy term babies.

**CONCLUSION** The results of our systematic review suggest that IFN-α does not significantly increase the risk of major malformation, miscarriage, stillbirth or preterm delivery above general population rates. It is also possible that IFN-α may have a protective effect against pregnancy loss in cases of ET.

**Database:** Medline

28. Successful outcome of a pregnancy with essential thrombocythaemia: A case report and literature review

**Author(s):** Nausheen A.; Syed A.

**Source:** BJOG: An International Journal of Obstetrics and Gynaecology; Jun 2012; vol. 119; p. 76

**Publication Date:** Jun 2012

**Publication Type(s):** Conference Abstract

**Abstract:** Essential thrombocythaemia is the commonest myeloproliferative disorder in women of child bearing age. A number of case reports have been described in literature but the optimum management of this disorder is still not established. Here we report a case of essential thrombocythaemia in pregnancy with good outcome for both mother and baby. Due to rarity of this disorder UKOSS is collecting data on myeloproliferative disorder in pregnancy. We feel that reporting this case will add to the current literature and aid decision making in care of these patients.

**Database:** EMBASE
29. How to manage essential thrombocythemia.
Author(s): Finazzi, G
Source: Leukemia; May 2012; vol. 26 (no. 5); p. 875-882
Publication Date: May 2012
Publication Type(s): Journal Article Review
PubMedID: 22051530
Available at Leukemia - from ProQuest (Hospital Premium Collection) - NHS Version
Available at Leukemia - from Unpaywall
Abstract: I use the hematological, morphological and molecular criteria recently established by the World Health Organization to diagnose essential thrombocythemia. In these patients, major causes of morbidity and mortality are represented by thrombosis and bleeding, whereas progression to myelofibrosis and transformation to acute leukemia are more rare. Myelosuppressive therapy can reduce the rate of vascular complications, but there is some concern about treatment-related toxicity. Therefore, I follow a risk-oriented therapeutic approach to avoid inappropriate exposure to cytotoxic drugs on one side or suboptimal treatment on the other. Established predictors of cardiovascular events are represented by older age and previous thrombosis, whereas recent data suggest a prognostic role for novel risk factors, including leukocytosis and JAK2V617F mutational status. There is no indication for therapeutic intervention in asymptomatic, low-risk patients, while I treat high-risk patients with hydroxyurea (HU) first. Other therapeutic options, such as interferon alpha or anagrelide, may find place in selected patients including those who are resistant or intolerant to HU. I follow a risk-oriented approach also for management of pregnancy. Low-risk women are given low-dose aspirin throughout pregnancy and prophylactic low-molecular-weight heparin (LMWH) post partum, whereas LMWH throughout pregnancy and/or interferon-alpha can be required in high-risk cases.
Database: Medline

Author(s): Cervantes, Francisco
Publication Date: 2011
Publication Type(s): Research Support, Non-u.s. Gov't Journal Article Review
PubMedID: 22160037
Available at Hematology. American Society of Hematology. Education Program - from HighWire - Free Full Text
Available at Hematology. American Society of Hematology. Education Program - from Unpaywall
Abstract: Essential thrombocythemia (ET) is a Philadelphia chromosome (Ph)-negative myeloproliferative neoplasm (MPN) characterized by thrombocytosis and megakaryocytic hyperplasia of the bone marrow, with presence of the JAK2 V617F mutation in 50%-60% of patients. ET evolves to myelofibrosis in a minority of cases, whereas transformation to acute leukemia is rare and increases in association with the use of certain therapies. Survival of ET patients does not substantially differ from that of the general population. However, important morbidity is derived from vascular complications, including thrombosis, microvascular disturbances, and bleeding. Because of this, treatment of ET must be aimed at preventing thrombosis and bleeding without increasing the risk of transformation of the disease. Patients are considered at high risk of
thrombosis if they are older than 60 years or have a previous history of thrombosis and at high risk of bleeding if platelet counts are > 1500 × 10(9)/L. Patients with low-risk ET are usually managed with low-dose aspirin, whereas treatment of high-risk ET is based on the use of cytoxic reductive therapy, with hydroxyurea as the drug of choice and IFN-α being reserved for young patients or pregnant women. For patients resistant or intolerant to hydroxyurea, anagrelide is recommended as second-line therapy. Strict control of coexistent cardiovascular risk factors is mandatory for all patients. The role in ET therapy of new drugs such as pegylated IFN or the JAK2 inhibitors is currently under investigation.

**Database:** Medline

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31. **Thrombocytosis in pregnancy - Is there any need for further evaluation?**

**Author(s):** Bergmann F.; Westrich V.; Czwalinna A.; Platzer C.; Hempel M.

**Source:** Hamostaseologie; 2011; vol. 31 (no. 1)

**Publication Date:** 2011

**Publication Type(s):** Conference Abstract

**Abstract:** Introduction: Most gynecologists are not aware of the potential abnormality of an elevated platelet count in pregnant women. Iron deficiency is a frequent cause of elevated platelet counts. However, a bone marrow abnormality should be considered, especially if the woman has additional symptoms like arterial/venous thrombosis, recurrent fetal loss. Essential thrombocytosis (ET) (Jak2-kinase mutation positive in 50-60%) is a rare disorder, it is more common in women and most patients are diagnosed before age 50. Patients and Methods: In the last 2 years we investigated 9 pregnant women and one non-pregnant women with an elevated platelet count >500/μl (normal range 150-400/μl) to rule out a pathology. Whole blood count, Ferritin levels and Jak2-mutation were investigated. In women with thrombosis or fetal loss acquired or inherited thrombophilia was ruled out. Results: 1/10 women had a splenectomy, 2/10 were diagnosed with iron deficiency, 2/10 no abnormality was detected. 2/10 were Jak2-mutation negative (bone marrow not investigated), however one women had recurrent thrombosis, one had 5 miscarriages (chromosomal abnormality excluded). In 3/10 women Jak2-mutation was detected and diagnosis of ET was established. One woman had an arterial thrombosis before pregnancy, she delivered a healthy child at term, receiving LMW heparin during pregnancy. One woman was infertile and is pregnant with twins (IVF) currently, treated with interferon and ASA. The third is planning to get pregnant - no treatment yet. Discussion: In pregnant women thrombocytosis needs further investigation, because women with ET have a 30% change for fetal loss and 8% suffer from maternal complications. Current treatment options and the literature will be reviewed.

**Database:** EMBASE
32. Essential thrombocythemia and pregnancy.

**Author(s):** Valera, Marie-Cécile; Parant, Olivier; Vayssiere, Christophe; Arnal, Jean-François; Payrastre, Bernard

**Source:** European journal of obstetrics, gynecology, and reproductive biology; Oct 2011; vol. 158 (no. 2); p. 141-147

**Publication Date:** Oct 2011

**Publication Type(s):** Journal Article Review

**PubMedID:** 21640467

**Abstract:** Essential thrombocythaemia (ET) is an acquired myeloproliferative neoplasm, characterised by persistent thrombocytosis and a tendency for either thrombosis or haemorrhage. Among myeloproliferative neoplasms, ET is the most prevalent in young women, which constitute a special group due to their childbearing potential. An increased risk of fetal and maternal complications has been demonstrated in patients with ET. The most common pregnancy-related complication is spontaneous abortion during the first trimester. Recurrent abortion, fetal growth restriction, stillbirth and placental abruption are less frequent. Maternal complications are relatively rare and essentially represented by thromboembolic and bleeding events. Here we summarize the literature describing pregnancy and its outcome in patients with ET and discuss some recommendations for the management of pregnancy.

**Database:** Medline

33. Myeloproliferative Disorders in Pregnancy

**Author(s):** Harrison C.N.; Robinson S.E.

**Source:** Hematology/Oncology Clinics of North America; Apr 2011; vol. 25 (no. 2); p. 261-275

**Publication Date:** Apr 2011

**Publication Type(s):** Review

**PubMedID:** 21444029

**Abstract:** This article reviews pregnancy outcome in women diagnosed with a myeloproliferative neoplasm (MPN), and discusses possible risk markers and the pathogenesis of poor pregnancy outcome. An outline of the key factors regarding the diagnosis and management of MPN in women of reproductive potential is followed by a description of the authors' management strategy for standard and high-risk pregnancy in MPN patients. © 2011 Elsevier Inc.

**Database:** EMBASE

**Author(s):** Palandri, Francesca; Polverelli, Nicola; Ottaviani, Emanuela; Castagnetti, Fausto; Baccarani, Michele; Vianelli, Nicola

**Source:** Haematologica; Jun 2010; vol. 95 (no. 6); p. 1038-1040

**Publication Date:** Jun 2010

**Publication Type(s):** Research Support, Non-u.s. Gov't Comparative Study Letter

**PubMedID:** 20081056

Available at Haematologica - from Europe PubMed Central - Open Access

**Database:** Medline

35. Aspirin in pregnant patients with essential thrombocythemia: a retrospective analysis of 129 pregnancies.

**Author(s):** Passamonti, F; Rumi, E; Randi, M L; Morra, E; Cazzola, M

**Source:** Journal of thrombosis and haemostasis : JTH; Feb 2010; vol. 8 (no. 2); p. 411-413

**Publication Date:** Feb 2010

**Publication Type(s):** Research Support, Non-u.s. Gov't Letter

**PubMedID:** 19912517

Available at Journal of thrombosis and haemostasis : JTH - from Wiley Online Library Science, Technology and Medicine Collection 2017

Available at Journal of thrombosis and haemostasis : JTH - from Unpaywall

**Database:** Medline


**Author(s):** Séror, Jérémy; Sentilhes, Loïc; Lefebvre-Lacoeuille, Céline; Marpeau, Loïc

**Source:** Fetal diagnosis and therapy; 2009; vol. 25 (no. 1); p. 136-140

**Publication Date:** 2009

**Publication Type(s):** Case Reports Journal Article Review

**PubMedID:** 19912590

Available at Fetal diagnosis and therapy - from ProQuest (Hospital Premium Collection) - NHS Version

**Abstract:** Described as a myeloproliferative disorder mainly affecting elderly women, recent reports now confirm the occurrence of essential thrombocythemia at younger ages, which questions treatment during pregnancy. We report a further case of uneventful full term pregnancy with the use of interferon-alpha for maternal essential thrombocythemia which suggests that interferon-alpha could be considered as an effective and safe treatment during pregnancy for women with essential thrombocythemia. Further studies are warranted to determine whether interferon-alpha is the optimal therapeutic option during pregnancy for this patient population.

**Database:** Medline
37. Outcome of 122 pregnancies in essential thrombocythemia patients: A report from the Italian registry.

**Author(s):** Melillo, Lorella; Tieghi, Alessia; Candoni, Anna; Radaelli, Franca; Ciancia, Rosanna; Specchia, Giorgina; Martino, Bruno; Scalzulli, Potito Rosario; Latagliata, Roberto; Palmieri, Fausto; Usala, Emilio; Valente, Daniela; Valvano, Maria Rosa; Cedrone, Michele; Comitini, Giuseppina; Martinelli, Vincenzo; Cascavilla, Nicola; Gugliotta, Luigi

**Source:** American journal of hematology; Oct 2009; vol. 84 (no. 10); p. 636-640

**Publication Date:** Oct 2009

**Publication Type(s):** Research Support, Non-u.s. Gov't Journal Article

**PubMedID:** 19705431

Abstract: Pregnancy is a high-risk event in women with essential thrombocythemia (ET). This observational study evaluated pregnancy outcome in ET patients focusing on the potential impact of aspirin (ASA) or interferon alpha (IFN) treatment during pregnancy. We retrospectively analyzed 122 pregnancies in 92 women consecutively observed in the last 10 years in 17 centers of the Italian thrombocythemia registry (RIT). The live birth rate was 75.4% (92/122 pregnancies). The risk of spontaneous abortion was 2.5-fold higher than in the control population (P < 0.01). ASA did not affect the live birth rate (71/93, 76.3% vs. 21/29, 72.4%, P = 0.67). However, IFN treatment during pregnancy was associated with a better outcome than was management without IFN (live births 19/20, 95% vs. 73/102, 71.6%, P = 0.025), and this finding was supported by multivariate analysis (OR: 0.10; 95% CI: 0.013-0.846, P = 0.034). The JAK2 V617F mutation was associated with a poorer outcome (fetal losses JAK2 V617F positive 9/25, 36% vs. wild type 2/24, 8.3%, P = 0.037), and this association was still significant after multivariate analysis (OR: 6.19; 95% CI: 1.17-32.61; P = 0.038). No outcome concordance between first and second pregnancies was found (P = 0.30). Maternal complications occurred in 8% of cases. In this retrospective study, in consecutively observed pregnant ET patients, IFN treatment was associated with a higher live birth rate, while ASA treatment was not. In addition, the JAK2 V617F mutation was confirmed to be an adverse prognostic factor.

**Database:** Medline

38. Essential thrombocythemia and pregnancy: Observations from recent studies and management recommendations.

**Author(s):** Tefferi, Ayalew; Passamonti, Francesco

**Source:** American journal of hematology; Oct 2009; vol. 84 (no. 10); p. 629-630

**Publication Date:** Oct 2009

**Publication Type(s):** Journal Article

**PubMedID:** 19731306

Abstract: Pregnancy is a high-risk event in women with essential thrombocythemia (ET). This observational study evaluated pregnancy outcome in ET patients focusing on the potential impact of aspirin (ASA) or interferon alpha (IFN) treatment during pregnancy. We retrospectively analyzed 122 pregnancies in 92 women consecutively observed in the last 10 years in 17 centers of the Italian thrombocythemia registry (RIT). The live birth rate was 75.4% (92/122 pregnancies). The risk of spontaneous abortion was 2.5-fold higher than in the control population (P < 0.01). ASA did not affect the live birth rate (71/93, 76.3% vs. 21/29, 72.4%, P = 0.67). However, IFN treatment during pregnancy was associated with a better outcome than was management without IFN (live births 19/20, 95% vs. 73/102, 71.6%, P = 0.025), and this finding was supported by multivariate analysis (OR: 0.10; 95% CI: 0.013-0.846, P = 0.034). The JAK2 V617F mutation was associated with a poorer outcome (fetal losses JAK2 V617F positive 9/25, 36% vs. wild type 2/24, 8.3%, P = 0.037), and this association was still significant after multivariate analysis (OR: 6.19; 95% CI: 1.17-32.61; P = 0.038). No outcome concordance between first and second pregnancies was found (P = 0.30). Maternal complications occurred in 8% of cases. In this retrospective study, in consecutively observed pregnant ET patients, IFN treatment was associated with a higher live birth rate, while ASA treatment was not. In addition, the JAK2 V617F mutation was confirmed to be an adverse prognostic factor.

**Database:** Medline

Author(s): Gangat, Naseema; Wolanskyj, Alexandra P; Schwager, Susan; Tefferi, Ayalew

Source: European journal of haematology; May 2009; vol. 82 (no. 5); p. 350-353

Publication Date: May 2009

Publication Type(s): Journal Article

PubMedID: 19243425

Available at European journal of haematology - from Wiley Online Library Science, Technology and Medicine Collection 2017

Abstract: An increased risk of pregnancy complications was recently reported in JAK2V617F-positive essential thrombocythemia (ET). In the current study of 63 pregnancies among 36 women with ET, we sought to appraise this association and identify other predictors of outcome. Overall outcome included 38 (60%) births and 20 (35%) first trimester spontaneous abortions. Among 36 first pregnancies, 22 (61%) resulted in live birth. Twelve of the 14 pregnancy losses occurred during the first trimester. Rate of pregnancy loss was 21% among 24 patients receiving aspirin therapy during the first trimester vs. 75% among 12 patients not receiving such treatment (P = 0.002). Pregnancy outcome was not influenced by platelet count, leukocyte count or presence of JAK2V617F; four pregnancy losses each were documented in 10 mutated and 10 unmutated patients. Among 17 second pregnancies, 12 (71%) resulted in live birth; these included eight from nine patients with successful and four from eight with unsuccessful first pregnancies (P = 0.07). Maternal complications were infrequent (11%): pre-eclampsia (n = 1), hematoma after Cesarean-section (n = 2) and post-partum hemorrhage (n = 1). This study suggests a salutary role for aspirin therapy in pregnant women with ET. Furthermore, the occurrence of a miscarriage in ET might be a marker for a similar event during subsequent pregnancies.

Database: Medline

Author(s): Griesshammer, Martin; Struve, Sabine; Barbui, Tiziano

Source: Blood reviews; Sep 2008; vol. 22 (no. 5); p. 235-245

Publication Date: Sep 2008

Publication Type(s): Research Support, Non-u.s. Gov't Journal Article Review

PubMedID: 18617299

Abstract: The management of pregnancy in Philadelphia negative chronic myeloproliferative disorders (CMPDs) is an increasingly frequent problem. In the literature, most pregnancies are reported for women with essential thrombocythemia (ET) with about 400 pregnancies in about 200 women. In ET, first trimester abortion is the most frequent complication occurring in about one third of pregnancies. Interestingly, the incidence of maternal complications is relatively low with 3% for major thromboembolic and 2% for major bleeding events. The presence of the Jak2 mutation seems to be an independent predictor of pregnancy complications. Pregnancies in ET should be stratified according to underlying risk factors in low, high and highest risk pregnancies. Women with low risk pregnancies are treated with low-dose aspirin, whereas women with high and higher risk pregnancies may benefit from low-dose aspirin plus interferon alpha +/- low molecular weight heparin throughout pregnancy and at least for six weeks post-partum. In polycythemia vera (PV) there is only very few information on pregnancy outcome with 36 pregnancies reported in the literature. According to these data pregnancy in PV is per se a high risk situation. Accordingly, all women with PV should be treated with low-dose aspirin. Some pregnant PV patients may benefit from a more intensive therapy including interferon alpha +/- low molecular weight heparin throughout pregnancy and at least for six weeks post-partum.

Database: Medline

41. Thrombocytosis, pregnancy, and regression toward mean

Author(s): Spivak J.L.

Source: Blood; Jul 2007; vol. 110 (no. 2); p. 472-473

Publication Date: Jul 2007

Publication Type(s): Note

Available at Blood - from HighWire - Free Full Text . com

Available at Blood - from Unpaywall

Database: EMBASE
42. Essential thrombocythemia.

**Author(s):** Brière, Jean B

**Source:** Orphanet journal of rare diseases; Jan 2007; vol. 2; p. 3

**Publication Date:** Jan 2007

**Publication Type(s):** Journal Article Review

**PubMedID:** 17210076

Available at Orphanet journal of rare diseases - from BioMed Central

Available at Orphanet journal of rare diseases - from Europe PubMed Central - Open Access

**Abstract:** Essential thrombocythemia (ET) is an acquired myeloproliferative disorder (MPD) characterized by a sustained elevation of platelet number with a tendency for thrombosis and hemorrhage. The prevalence in the general population is approximately 30/100,000. The median age at diagnosis is 65 to 70 years, but the disease may occur at any age. The female to male ratio is about 2:1. The clinical picture is dominated by a predisposition to vascular occlusive events (involving the cerebrovascular, coronary and peripheral circulation) and hemorrhages. Some patients with ET are asymptomatic, others may experience vasomotor (headaches, visual disturbances, lightheadedness, atypical chest pain, distal paresthesias, erythromelalgia), thrombotic, or hemorrhagic disturbances. Arterial and venous thromboses, as well as platelet-mediated transient occlusions of the microcirculation and bleeding, represent the main risks for ET patients. Thromboses of large arteries represent a major cause of mortality associated with ET or can induce severe neurological, cardiac or peripheral artery manifestations. Acute leukemia or myelodysplasia represent only rare and frequently later-onset events. The molecular pathogenesis of ET, which leads to the overproduction of mature blood cells, is similar to that found in other clonal MPDs such as chronic myeloid leukemia, polycythemia vera and myelofibrosis with myeloid metaplasia of the spleen. Polycythemia vera, myelofibrosis with myeloid metaplasia of the spleen and ET are generally associated under the common denomination of Philadelphia (Ph)-negative MPDs. Despite the recent identification of the JAK2 V617F mutation in a subset of patients with Ph-negative MPDs, the detailed pathogenetic mechanism is still a matter of discussion. Therapeutic interventions in ET are limited to decisions concerning the introduction of anti-aggregation therapy and/or starting platelet cytoreduction. The therapeutic value of hydroxyurea and aspirin in high risk patients has been supported by controlled studies. Avoiding thromboreduction or opting for anagrelide to postpone the long-term side effects of hydroxyurea in young or low risk patients represent alternative options. Life expectancy is almost normal and similar to that of a healthy population matched by age and sex.

**Database:** Medline
43. Interferon-alfa treatment of essential thrombocythemia during pregnancy

Author(s): Iwashita T.; Fujitani M.; Yamamoto Y.; Katsurada T.; Yoshida Y.

Source: Internal Medicine; Nov 2006; vol. 45 (no. 20); p. 1161-1164

Publication Date: Nov 2006

Publication Type(s): Article

PubMedID: 17106162

Available at Internal medicine (Tokyo, Japan) - from Unpaywall

Abstract: We report on 2 successful pregnancies in a young woman who has essential thrombocythemia. The platelet count remained well controlled with interferon-alfa administration together with acetylsalicylic acid in each pregnancy. The present case and the published series suggest that close monitoring of the platelet count is crucial and that interferon may be the preferred therapeutic option in the management of pregnancy in patients with essential thrombocythemia. © 2006 The Japanese Society of Internal Medicine.

Database: EMBASE

44. Essential thrombocythemia/polycythemia vera and pregnancy: The need for an observational study in Europe

Author(s): Griesshammer M.; Struve S.; Harrison C.M.

Source: Seminars in Thrombosis and Hemostasis; Jun 2006; vol. 32 (no. 4); p. 422-429

Publication Date: Jun 2006

Publication Type(s): Article

PubMedID: 16810618

Available at Seminars in Thrombosis and Hemostasis - from Unpaywall

Abstract: The management of pregnant patients with essential thrombocythemia (ET) and polycythemia vera (PV) may be problematic. In the literature there are ~300 cases of pregnancies reported in ET and less than 50 pregnancies reported in PV. To reduce the effect of reporting bias, we selected articles with either >10 pregnancies or at least six patients, and here report on the outcome of 195 pregnancies in ET and 36 pregnancies in PV patients. The live birth rate was ~60% in ET and 58% in PV. Spontaneous abortion during the first trimester was the most frequent fetal complication, occurring in 31% of ET pregnancies and in 22% of PV pregnancies, respectively. Major maternal complications were more frequent in PV compared with ET (44.4 versus 7.7%). Treatment with low-dose aspirin during pregnancy in ET seemed to reduce complications and also seemed beneficial during pregnancy in PV. In high-risk pregnancies, the additional use of low molecular weight heparin and/or interferon alpha should be considered. This article also describes a registry for an observational study concerning pregnancy in chronic Philadelphia chromosome-negative myeloproliferative disorders within the European LeukemiaNet. A potential management algorithm for pregnancies in ET or PV is also provided. Copyright © 2006 by Thieme Medical Publishers, Inc.

Database: EMBASE
45. Pregnancy and its management in the Philadelphia negative myeloproliferative diseases

Author(s): Harrison C.
Source: British Journal of Haematology; 2005; vol. 129 (no. 3); p. 293-306
Publication Date: 2005
Publication Type(s): Review
PubMedID: 15842653
Available at British journal of haematology - from Wiley Online Library Science, Technology and Medicine Collection 2017
Available at British journal of haematology - from Unpaywall

Abstract: The myeloproliferative diseases (MPDs) present several therapeutic challenges in patients of childbearing potential. The most extensive literature exists for patients with essential thrombocythaemia, with over 200 pregnancies reported in retrospective case series. Yet there is conflicting data in relation to predicting pregnancy outcome and optimal management strategy. Pregnancy is less frequently reported for polycythaemia vera and myelofibrosis. There is a need for collaboration to further our knowledge in this field. Here, the literature is reviewed in detail and experience of different therapeutic strategies in pregnancy discussed. There is increasing understanding about the pathogenesis of placental dysfunction in inherited thrombophilia and antiphospholipid antibody syndrome pregnancy outcomes in these conditions parallel those reported for MPDs. Furthermore several large studies have influenced pregnancy management in these conditions and, whilst not directly applicable to MPDs, this data have potential to inform treatment protocols. This data are reviewed and a personal management strategy for pregnancy in MPD pro-Dosed. © 2005 Blackwell Publishing Ltd.

Database: EMBASE

46. Pregnancy in essential thrombocythaemia: experience with 40 pregnancies.

Author(s): Niittyvuopio, Riitta; Juvonen, Eeva; Kaaja, Risto; Oksanen, Kalevi; Hallman, Heikki; Timonen, Timo; Ruutu, Tapani
Source: European journal of haematology; Dec 2004; vol. 73 (no. 6); p. 431-436
Publication Date: Dec 2004
Publication Type(s): Journal Article Review
PubMedID: 15522066
Available at European journal of haematology - from Wiley Online Library Science, Technology and Medicine Collection 2017

Abstract: In this study, the course of 40 pregnancies in 16 women with essential thrombocythaemia (ET) was analysed retrospectively. Of the pregnancies, 45% were complicated, 55% uncomplicated, and 62% resulted in live birth. The most common complication was spontaneous abortion during the first trimester seen in 33% of all pregnancies and comprising 72% of all complications. Two intrauterine foetal deaths occurred at weeks 22 and 28. Three pregnancies were complicated by eclampsia or pre-eclampsia. Nine of 16 women with 29 pregnancies had at least one complicated pregnancy. In seven of 16 women, all 11 pregnancies were uneventful. The non-pregnancy-related symptoms of ET or the platelet count before conception or during pregnancy did not correlate with the risk of pregnancy complications. Treatment with low-dose acetylsalicylic acid (ASA) alone during pregnancy or platelet-lowering drugs before or during pregnancy reduced the risk of complications.

Database: Medline
47. Interferon alfa treatment for pregnant women affected by essential thrombocythemia: case reports and a review.

Author(s): Martinelli, Pasquale; Martinelli, Vincenzo; Agangi, Annalisa; Maruotti, Giuseppe Maria; Paladini, Dario; Ciancia, Rosanna; Rotoli, Bruno

Source: American journal of obstetrics and gynecology; Dec 2004; vol. 191 (no. 6); p. 2016-2020

Publication Date: Dec 2004

Publication Type(s): Case Reports Journal Article Review

PubMedID: 15592285

Abstract: OBJECTIVES In the past essential thrombocythemia was considered a disease of the elderly. At present, the number of young people suffering from this disease is growing, with a slightly higher frequency in females. We investigated the effects of interferon alfa therapy in these patients. STUDY DESIGN We describe 9 pregnancies in 4 women affected by essential thrombocythemia. RESULTS Four pregnancies were carried out without interferon alfa therapy, and resulted in 2 intrauterine deaths, 1 spontaneous abortion, and 1 neonatal death. Interferon alfa was given during another 5 pregnancies; among them, 2 ended in preterm deliveries with normal infants, and 3 in full-term deliveries. The literature is reviewed. CONCLUSION Our cases and published series suggest that fetal outcome is improved by therapy, and that interferon alfa may be the best therapeutic option.

Database: Medline

48. Practice guidelines for the therapy of essential thrombocythemia. A statement from the Italian Society of Hematology, the Italian Society of Experimental Hematology and the Italian Group for Bone Marrow Transplantation.

Author(s): Barbui, Tiziano; Barosi, Giovanni; Grossi, Alberto; Gugliotta, Luigi; Liberato, Lucio N; Marchetti, Monia; Mazzucconi, Maria Gabriella; Rodeghiero, Francesco; Tura, Sante

Source: Haematologica; Feb 2004; vol. 89 (no. 2); p. 215-232

Publication Date: Feb 2004

Publication Type(s): Practice Guideline Guideline Journal Article Consensus Development Conference Review

PubMedID: 15003898

Abstract: BACKGROUND AND OBJECTIVES The Italian Society of Hematology (SIE) and the two affiliated Societies (SIES and GITMO) commissioned a project to develop guidelines for the therapy of essential thrombocythemia (ET) using evidence-based knowledge and consensus formation techniques. DESIGN AND METHODS Key questions on the optimal management of ET patients were formulated by an Advisory Council (AC) and approved by an Expert Panel (EP) composed of 7 senior hematologists. The AC systematically reviewed the published literature from 1980 to August 2002, and articles were graded according to their internal validity and quality. Using the Delphi technique, the EP was asked to answer the key questions according to the available evidence. From September 2002 to March 2003, four Consensus Conferences were held in accordance with the Nominal Group Technique with the goal of solving residual disagreement on recommendations. RESULTS The EP provided recommendations on when to start platelet-lowering therapy, the most appropriate platelet-lowering agent, the use of anti-platelet therapy, and the management of women in childbearing age and of pregnant women. INTERPRETATION AND CONCLUSIONS By using evidence and consensus, recommendations for the treatment of key problems in ET have been issued. Statements are graded according to the strength of the supporting evidence and uncertainty is explicitly declared.

Database: Medline
49. Thrombocythaemia and pregnancy.

**Author(s):** Elliott, Michelle A; Tefferi, Ayalew

**Source:** Best practice & research. Clinical haematology; Jun 2003; vol. 16 (no. 2); p. 227-242

**Publication Date:** Jun 2003

**Publication Type(s):** Journal Article Review

**PubMedID:** 12763489

**Abstract:** The diagnosis of essential thrombocythaemia (ET) is considered when all causes of reactive (secondary) thrombocytosis have been excluded in a patient with a persistent thrombocytosis. Among the chronic myeloproliferative disorders, ET has the highest proportion of young as well as female patients affected. As such, the optimal management of ET during pregnancy becomes an important issue. Although ET is not a contraindication to childbearing, the risks to both the fetus and mother are increased, and would-be-parents should be counselled regarding these. Obstetric complications occurring at higher than expected rates include spontaneous abortion in the first trimester. Maternal complications, both haemorrhagic and thrombotic, are reported relatively infrequently. It is generally difficult to make specific management recommendations because of the relative rarity of the association between ET and pregnancy, which has precluded adequately powered controlled trials. In this communication, we review our own experience as well as that of other investigators on the subject matter and provide management guidelines that are based on best available information.

**Database:** Medline


**Author(s):** Vantroyen, B; Vanstraelen, D

**Source:** Acta haematologica; 2002; vol. 107 (no. 3); p. 158-169

**Publication Date:** 2002

**Publication Type(s):** Journal Article Review

**PubMedID:** 11978937

**Abstract:** It is advisable to treat essential thrombocytemia (ET) during pregnancy, because elevated platelet counts can lead to maternal and fetal complications. In order to establish which therapy is more favorable, we undertook a review of the literature. In addition to our own case, we found 27 reports which described 75 cases with 143 pregnancies. We discussed the complications of ET during pregnancy and postpartum, fetal outcome and the therapeutic strategies. Considering the clear risk of complications during pregnancy -- especially the occurrence of spontaneous abortion in the first trimester -- and the risk of intrauterine fetal death, we believe all patients should at least be treated with aspirin unless there is a contraindication. Platelet reduction with interferon-alpha (IFN-alpha) might be able to further reduce the complications of ET during pregnancy and to improve fetal outcome (data from 14 patients). After treatment with IFN-alpha, sufficient numbers of umbilical cord blood stem cells can be collected.

**Database:** Medline
51. Platelet disorders in pregnancy
Author(s): Burrows R.F.
Source: Current Opinion in Obstetrics and Gynecology; 2001; vol. 13 (no. 2); p. 115-119
Publication Date: 2001
Publication Type(s): Review
PubMedID: 11315863
Available at Current opinion in obstetrics & gynecology - from Ovid (LWW Total Access Collection 2015 - Q1 with Neurology)
Abstract: Before the advent of automated cell counters, thrombocytopenia was looked for and identified after a clinical question. With automation, thrombocytopenia in pregnancy was commonly found, and with its commonality its clinical meaning went out of focus. For the majority of women, thrombocytopenia is benign and, as a marker of fetal thrombocytopenia, the value of maternal thrombocytopenia is almost non-existent. © 2001 Lippincott Williams & Wilkins.
Database: EMBASE

52. A single institutional experience with 43 pregnancies in essential thrombocythemia
Author(s): Wright C.A.; Tefferi A.
Source: European Journal of Haematology; 2001; vol. 66 (no. 3); p. 152-159
Publication Date: 2001
Publication Type(s): Article
PubMedID: 11350483
Available at European journal of haematology - from Wiley Online Library Science, Technology and Medicine Collection 2017
Abstract: Objectives: We describe the periconception circumstances and outcome of 43 consecutive pregnancies in an unselected group of young women with essential thrombocythemia (ET). Patients and methods: We retrospectively studied 74 consecutive cases of young women with ET seen at our institution, among whom 43 pregnancies occurred in 20 patients. Results: Of the 43 pregnancies, 22 (51%) were successful (21 term and 1 preterm live births) and 21 (49%) ended in miscarriages (1 ectopic pregnancy, 2 elective abortions, 16 first-trimester spontaneous abortions, 1 stillbirth at 22 wk, and 1 abruptio placentae at 33 wk). Management of ET at the time of conception included either no specific therapy (16 cases) or the use of aspirin alone (24 cases), a cytoreductive agent (2 cases), or heparin (1 case). There were no significant differences with respect to platelet count or the effect of treatment with aspirin, either at the time of conception or during the first trimester, among cases of successful pregnancies (22), all miscarriages (21), or first-trimester spontaneous abortions (16). The findings were similar when the analysis was restricted to only first-time pregnancies. In patients with multiple pregnancies, the outcome of a subsequent pregnancy was not predicted by the outcome of the first. In general, in successful cases the last two trimesters were mostly uneventful, with healthy offspring being reported in all cases. Conclusions: Pregnant patients with ET have an increased risk of first-trimester abortion which is not predictable by preconception platelet count or aspirin therapy, in addition, our experience does not support the use of prophylactic platelet apheresis during delivery.
Database: EMBASE
Primary myelofibrosis with thrombocytosis in pregnancy: A case report

Author(s): Bozanovic T.; Cvetkovic M.; Ljubic A.; Kesic V.; Dukanac J.; Ciric R.; Gotic M.

Source: Prenatal and Neonatal Medicine; 2000; vol. 5 (no. 3); p. 189-192

Publication Date: 2000

Publication Type(s): Article

Abstract: Primary myelofibrosis is a myeloproliferative disease. Some 12% of patients with primary myelofibrosis develop thrombocytosis. Thrombocytosis in pregnancy leads to higher rates of spontaneous abortion, premature delivery, intrauterine growth restriction and fetal death. The aim of the authors is to present the course, therapy and outcome of pregnancy in a patient with primary myelofibrosis and thrombocytosis. Myelosuppressive therapy decreases the incidence of thrombosis, but is contraindicated in pregnancy. Owing to this impossibility of treatment, pregnancies complicated by thrombocytosis have unfavorable outcomes. Based on data showing a favorable pregnancy outcome after administration of interferon-alpha in essential thrombocythaemia, it was decided to use the same drug for the first time in a pregnant patient with primary myelofibrosis and thrombocytosis. Close follow-up and continuous adjustment of the therapy resulted in a normal course and outcome of the pregnancy. To the best of our knowledge, this is the first report of successful treatment with interferon-alpha in a pregnant patient with primary myelofibrosis and thrombocytosis.

Database: EMBASE

Pregnancy in essential thrombocythaemia: treatment and outcome of 17 pregnancies.

Author(s): Bangerter, M; Guthner, C; Beneke, H; Hildebrand, A; Grunewald, M; Griesshammer, M

Source: European journal of haematology; Sep 2000; vol. 65 (no. 3); p. 165-169

Publication Date: Sep 2000

Publication Type(s): Journal Article

PubMedID: 11007051

Available at European journal of haematology - from Wiley Online Library Science, Technology and Medicine Collection 2017

Abstract: OBJECTIVE To evaluate treatment and outcome of 17 pregnancies in nine patients with essential thrombocythaemia (ET) seen at our institution from 1988 to 1998. METHODS Treatment and outcome of 17 pregnancies in nine ET patients were retrospectively analyzed. RESULTS Seventeen pregnancies in nine patients with ET resulted in 11 (65%) live births and ended in six (35%) spontaneous abortions. Abortion could not be predicted from ET-associated complications before (p=0.23) or during (p=0.39) pregnancy. Maternal complications occurred during six pregnancies (35%): Three major bleedings in two patients with an acquired von Willebrand disease and two minor bleedings in patients treated with low-dose acetylsalicylic acid (ASA) were observed during pregnancy or at term; one patient suffered from transient visual loss while pausing low-dose ASA. Platelet counts prior to pregnancy were significantly higher as compared to the platelet nadir observed during pregnancy (p=0.0017). Postpartum clinical course was uneventful in all patients. No specific treatment was given during 11 pregnancies. Six women received low-dose ASA during pregnancy followed by low-molecular-weight heparin until the end of the sixth week postpartum in five cases. This treatment was correlated with a favourable outcome (live birth versus abortion) when compared to no treatment (p=0.04). CONCLUSION Pregnancy in ET can be complicated by first trimester abortion and/or maternal haemorrhage. Our limited observation suggest a positive impact of low-dose ASA during pregnancy followed by low-molecular-weight heparin postpartum on pregnancy outcome in ET; nevertheless, confirmation by prospective documentation is mandatory.

**Author(s):** Cincotta, R; Higgins, J R; Tippett, C; Gallery, E; North, R; McMahon, L P; Brennecke, S P

**Source:** The Australian & New Zealand journal of obstetrics & gynaecology; Feb 2000; vol. 40 (no. 1); p. 33-37

**Publication Date:** Feb 2000

**Publication Type(s):** Journal Article

**PubMedID:** 10870776


**Abstract:** Essential thrombocythaemia is a rare myeloproliferative disorder that often presents with haemorrhagic or thrombotic complications. It may be detected incidentally in an asymptomatic younger adult and there are only a few case reports of essential thrombocythaemia in pregnant women. The risks posed by essential thrombocythaemia during pregnancy and its optimal management are uncertain. To determine if there is increased incidence of obstetric complications seen in women who have essential thrombocythaemia, we collected a large case series from a number of tertiary obstetric units in Australia and New Zealand. There were 30 pregnancies in 12 women who had essential thrombocythaemia. There were 17 live births (57%), 7 stillbirths (23%), 5 miscarriages (17%) and 1 ectopic (3%). Five pregnancies were complicated by placental abruption. When the outcomes of those women who received treatment with aspirin or interferon were compared to those that did not receive any treatment, there was a trend towards a higher livebirth rate (79% v. 38%, p = 0.06). Seven women were treated with aspirin and 5 had successful outcomes with no fetal complications. Four women were treated with alpha-interferon which reduced their platelet counts and all had successful outcomes with no fetal complication. These findings suggest that there is a high incidence of miscarriage, stillbirth and abruption in women with essential thrombocythaemia. Their pregnancies should be carefully monitored. Treatment with low dose aspirin and/or the use of alpha-interferon may be associated with an improved pregnancy outcome.

**Database:** Medline
56. Essential thrombocythemia in young adults: major thrombotic complications and complications during pregnancy—a follow-up study in 68 patients.

Author(s): Randi, M L; Rossi, C; Fabris, F; Girolami, A

Source: Clinical and applied thrombosis/hemostasis : official journal of the International Academy of Clinical and Applied Thrombosis/Hemostasis; Jan 2000; vol. 6 (no. 1); p. 31-35

Publication Date: Jan 2000

Publication Type(s): Research Support, Non-u.s. Gov't Journal Article

PubMedID: 10726046

Abstract: OBJECTIVES Although essential thrombocythemia (ET) is usually primarily considered a disorder of middle age, it has been observed in children and young adults. However, the real risk for thrombosis in these patients has not been clearly established. DESIGN Prospective analysis of consecutive patients younger than 40 at the time of the diagnosis of ET and followed in our department between 1980 and 1998. SUBJECTS Sixty-eight patients (28 males and 40 females, median follow-up 99.14 months) affected by ET diagnosed in agreement with the Polycythemia Vera Study Group criteria. INTERVENTIONS Asymptomatic ET patients were not treated. In contrast, patients with associated atherosclerotic risk factors, microvascular disturbances, or a previous major arterial thrombosis were given acetyl salicylic acid (ASA 100 mg/day). Only patients with major thrombotic complications and a platelet count > 1,000 x 10^9/L received cytoreductive therapy. OUTCOME MEASURES (1) to evaluate thrombotic complications in young patients with ET, (2) to relate thrombotic risk to the presence of general atherosclerotic risk factors, and (3) to adopt treatment, and (4) to report the outcome of the pregnancies monitored in our population. RESULTS Fifteen patients had major thrombosis, 11 of which were the presenting features of ET. No rethrombosis has been observed. Only one patient with thrombotic complications was under efficient treatment. Atherosclerotic risk factors are more common in patients with major arterial thrombosis than in asymptomatic subjects. Thirteen normal babies were delivered out of 16 pregnancies, 6 of the pregnant women were on ASA therapy. CONCLUSIONS Most thrombosis in young ET patients occurred at the time of the diagnosis, and venous thrombotic events represent one-third of total thrombosis. Cardiovascular risk factors seem to be concurrent stimuli for arterial thrombosis in ET. The thrombotic complication rate was 2.6/100 patients/year ASA reduces microvascular disturbances, thrombosis, and rethrombosis and possibly reduces obstetric complications in women with ET.

Database: Medline
57. Fertility, pregnancy and the management of myeloproliferative disorders

**Author(s):** Griesshammer M.; Bergmann L.; Pearson T.

**Source:** Bailliere's Clinical Haematology; 1998; vol. 11 (no. 4); p. 859-874

**Publication Date:** 1998

**Publication Type(s):** Review

**PubMedID:** 10640221

**Abstract:** The management of pregnant patients with chronic myeloproliferative disorders (MPD) is a difficult problem. Patients with essential thrombocythaemia (ET), and, less frequently, those with chronic myeloid leukaemia (CML) or polycythaemia vera (PV), present at a childbearing age. Pregnancy itself does not appear to affect adversely the natural course and prognosis of the MPD. However, fertility might be reduced, and an adverse outcome of pregnancy due to thrombotic or bleeding complications is a matter of concern. In ET, first-trimester abortion is the most frequent complication but increased perinatal mortality and premature delivery are also observed. Placental infarction due to thrombosis seems to be the most consistent event. Maternal thrombotic or haemorrhagic complications are rare but are more common than seen in normal pregnancy. The outcome of pregnancy seems to be positively influenced by aspirin, at least in some cases. The value of cytoreduction and/or heparin prophylaxis has not been established but may have a role in selected cases. In CML, the potential adverse effects of hyperleukocytosis, and sometimes thrombocytosis, generally make myelosuppressive treatment essential. In PV, the number of reported pregnancies is low. Maintaining the PCV below 0.45 is of the utmost importance relating to the outcome of pregnancy. Although cytoreductive drugs should generally be avoided, if possible, until at least after the first trimester of pregnancy, interferon-or seems to be the drug of choice when myelosuppression is indicated. In summary, the available information about pregnancy occurring during the course of an MPD indicates that successful management of pregnancy is possible. However, optimal management of these patients is poorly defined and agreed protocols are not available. In view of these problems, it is timely to consider the establishment of a national or European registry to monitor prospectively the management offered to pregnant women found to have an MPD.

**Database:** EMBASE

58. Pregnancy in a patient with essential thrombocytoysis

**Author(s):** Rossi G.; Appiano G.; Caremani M.; Lapini L.

**Source:** Clinical and Experimental Obstetrics and Gynecology; 1997; vol. 24 (no. 2); p. 114-115

**Publication Date:** 1997

**Publication Type(s):** Article

**PubMedID:** 9342481

**Abstract:** An insurgent case of pregnancy in a patient in whom essential thrombocytoysis was diagnosed five years earlier is described. Pregnancy was confirmed and therapy with platelet aggregation inhibitor was introduced. The pregnancy reached full term notwithstanding a positive result of the 'Triple Test' during the 15th week of gestation. A histology exam of the placenta revealed an ischemic lesion. We retain that platelet aggregation inhibitor therapy remains an important aid in eliminating the risk of thrombosis determined by the presence of two conditions that are predisposed to these risks, such as pregnancy and essential thrombocytoysis.

**Database:** EMBASE
59. Essential thrombocythemia during pregnancy.

Author(s): Eliyahu, S; Shalev, E

Source: Obstetrical & gynecological survey; Apr 1997; vol. 52 (no. 4); p. 243-247

Publication Date: Apr 1997

Publication Type(s): Case Reports Journal Article Review

PubMedID: 9095490

Available at Obstetrical & gynecological survey - from Ovid (LWW Total Access Collection 2015 - Q1 with Neurology)

Abstract: Essential thrombocythemia is a myeloproliferative disorder characterized by a persistent increase in the platelet count. The disease occurs more often in late middle age, but reports confirm the possibility of earlier occurrence. When the disease affects women during pregnancy, an adverse pregnancy outcome is possible: abortion, intrauterine fetal death, abruptio placentae, intrauterine growth retardation, and premature delivery. The possibility of thrombosis formation in this disease encouraged many physicians to use various treatments aimed at lowering the platelet count. Treatments used during pregnancy include acetylsalicylic acid, dipyridamole, heparin, and plateletpheresis. This article describes pregnancy outcomes and reviews the suggested treatments.

Database: Medline

60. Treatment of essential thrombocythemia during pregnancy with interferon- alpha

Author(s): Delage R.; Demers C.; Cantin G.; Roy J.

Source: Obstetrics and Gynecology; 1996; vol. 87 (no. 5); p. 814-817

Publication Date: 1996

Publication Type(s): Article

PubMedID: 8677098

Available at Obstetrics and Gynecology - from Ovid (LWW Total Access Collection 2015 - Q1 with Neurology)

Abstract: Background: Only a few cases of essential thrombocythemia in pregnant women have been reported, and the management of this myeloproliferative disorder during pregnancy remains uncertain. We report a successful pregnancy in a patient who had essential thrombocythemia and who was treated with interferon-alpha, and we review the literature for the outcome of similar patients. Case: A 32-year-old woman, gravida 4, para 3, aborta 0, presented at 18 weeks' gestation with two episodes of amaurosis fugax and an elevated platelet count of 2300 x 109/L. The initiation of interferon-alpha led to a progressive fall of the platelet level, with no occurrence of thrombotic or hemorrhagic manifestations. Serial ultrasound examinations revealed normal fetal and placental development. The patient was delivered of a male infant at 37 weeks. Both child and placenta were normal on examination. Conclusion: Our case and the current available data suggest that interferon-alpha may be the best therapeutic option for pregnant patients with essential thrombocythemia in whom myelosuppression is required.

Database: EMBASE
61. Essential thrombocythemia and pregnancy.

**Author(s):** Griesshammer, M; Heimpel, H; Pearson, T C

**Source:** Leukemia & lymphoma; Sep 1996; vol. 22

**Publication Date:** Sep 1996

**Publication Type(s):** Journal Article Review

**PubMedID:** 8951773

**Abstract:** A review of the literature disclosed 106 pregnancies (preg.) in 57 women with essential thrombocythemia (ET). The success rate (baby alive) was 57% (60 live births/106 preg.), the rate of miscarriage 43% (46 miscarriages/106 preg.). The most frequent complication was spontaneous abortion during the first trimester in 36% (38 abortions/106 preg.). Other complications such as intrauterine death and stillbirth after the 28th week, which occurred in 5% (7/106), premature delivery in 8% (8/106), pre-eclampsia in 4% (4/106), and fetal growth retardation in 4% (4/106) were rarer events. Placental infarction due to thrombosis seems to be the most consistent pathological event as far as the fetus is concerned. Maternal hemorrhage occurred in 4% (3 minor and 1 major bleeding) and only 2 minor maternal thrombotic episodes have been observed. Interestingly, a decline in platelet count has been observed in 14 women and was associated with a successful preg, in 13/14 cases (93%). Aspirin (ASS) was the most frequently used drug in 47 of 93 recorded cases (51%). In 16 evaluable women treated with ASS the live birth rate was higher (12/16 preg., 75%) than for 21 untreated women (9/21 preg., 43%). In 5 cases interferon alpha (IFN) has been used successfully. In summary, 57% of women with ET had a live birth, maternal complications happened in 6%. Promising treatment modalities might be ASS and IFN. However, no definitive answer can be given on the ideal management for women with ET during pregnancy. A European register should be set up.

**Database:** Medline

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62. Primary thrombocythemia and pregnancy: treatment and outcome in fifteen cases.

**Author(s):** Pagliaro, P; Arrigoni, L; Muggiasca, M L; Poggio, M; Russo, U; Rossi, E

**Source:** American journal of hematology; Sep 1996; vol. 53 (no. 1); p. 6-10

**Publication Date:** Sep 1996

**Publication Type(s):** Journal Article

**PubMedID:** 8813089

**Available at:** American journal of hematology - from Wiley Online Library Science, Technology and Medicine Collection 2017

**Available at:** American journal of hematology - from Unpaywall

**Abstract:** Pregnancy in patients with primary thrombocythemia (PT) is reported to be often complicated by recurrent abortion and fetal growth retardation. Fifteen pregnancies in nine patients with PT are reported. Nine pregnancies had a good outcome, with the birth of a healthy infant. There were two spontaneous abortions and three intrauterine deaths. One pregnancy was electively terminated after extensive thrombosis in the splanchnic district requiring surgical entero-resection. In five pregnancies the mother received no treatment; in ten pregnancies acetylsalicylic acid (ASA) was prescribed to the mother as soon as she was found pregnant, subcutaneous heparin was added from the middle trimester in seven cases. In patients treated with ASA and subcutaneous heparin pregnancies had a good outcome. Administration of ASA and heparin during pregnancy appears to improve the outcome in patients with PT and can prevent severe maternal complications, but requires close monitoring.

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