

# Essential (or primary) thrombocytopenia (ET) (short version)

Recommendations from the society for diagnosis and therapy of haematological and oncological diseases

## **Publisher**

DGHO Deutsche Gesellschaft für Hämatologie und  
Medizinische Onkologie e.V.  
Bauhofstr. 12  
D-10117 Berlin

Executive chairman: Prof. Dr. med. Andreas Hochhaus

Phone: +49 (0)30 27 87 60 89 - 0

[info@dgho.de](mailto:info@dgho.de)

[www.dgho.de](http://www.dgho.de)

## **Contact person**

Prof. Dr. med. Bernhard Wörmann  
Medical superintendent

## **Source**

[www.onkopedia-guidelines.info](http://www.onkopedia-guidelines.info)

The information of the DGHO Onkopedia Web Site is not intended or implied to be a substitute for professional medical advice or medical care. The advice of a medical professional should always be sought prior to commencing any form of medical treatment. To this end, all component information contained within the web site is done so for solely educational purposes. DGHO Deutsche Gesellschaft für Hämatologie und Onkologie and all of its staff, agents and members disclaim any and all warranties and representations with regards to the information contained on the DGHO Web Site. This includes any implied warranties and conditions that may be derived from the aforementioned web site information.

# Table of contents

<b>1 Summary .....</b>	<b>2</b>
<b>2 Therapy .....</b>	<b>2</b>
<b>15 Authors' Affiliations.....</b>	<b>3</b>
<b>16 Disclosures .....</b>	<b>4</b>

# Essential (or primary) thrombocythemia (ET) (short version)

**Date of document:** August 2021

**Compliance rules:**

- [Guideline](#)
- [Conflict of interests](#)

**Authors:** Petro E. Petrides, Gabriela M. Baerlocher, Konstanze Döhner, Heinz Gisslinger, Martin Grieshammer, Steffen Koschmieder, Juliana Schwaab, Eva Lengfelder

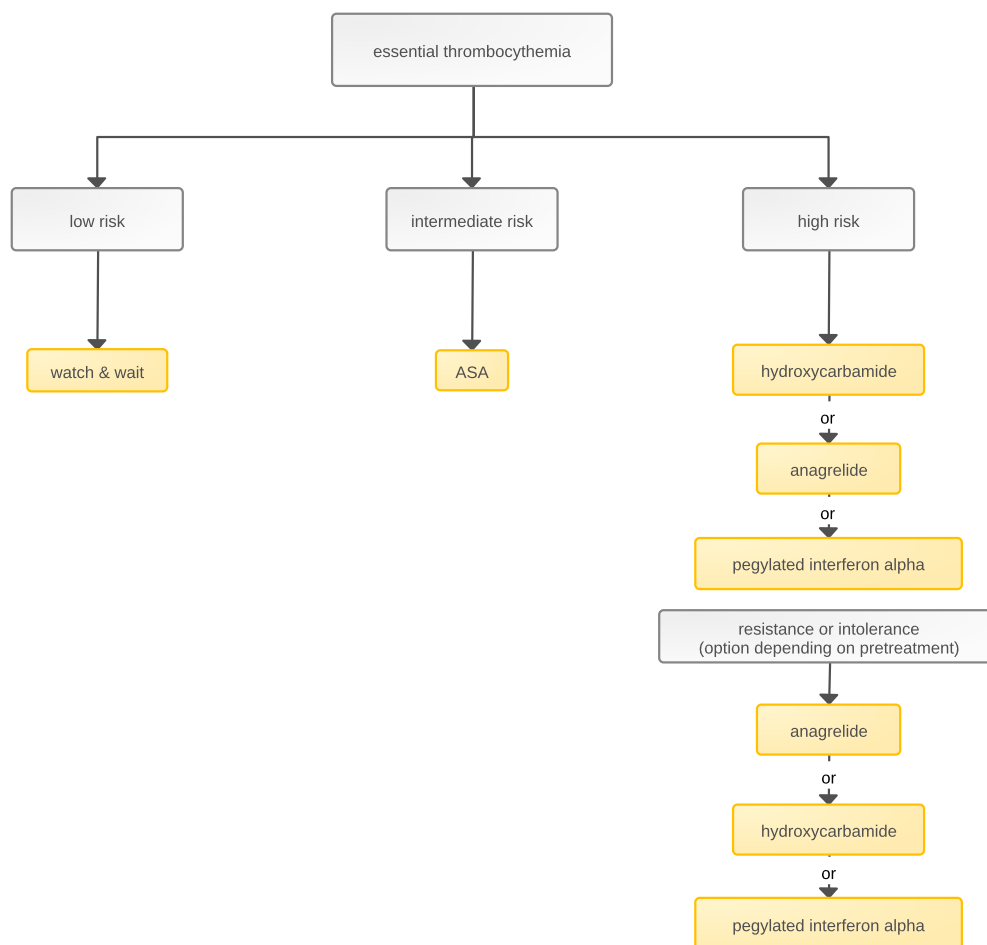
## 1 Summary

Essential (or primary) thrombocythemia (ET or PT) is a chronic myeloproliferative neoplasia (MPN) (formerly chronic myeloproliferative disease, CMPE) characterized by a persistent increase in platelet count. On the basis of molecular biological analyses (mutations in the *JAK2*, *CALR*, *MPL genes*), different subtypes can be distinguished. Typical disease complications are thrombosis and bleeding. Inhibitors of platelet aggregation and cytoreductive drugs are available for primary and secondary prevention of these complications. In a proportion of ET cases, the disease progresses to polycythaemia vera (PV), post-ET myelofibrosis or myelodysplastic syndrome (MDS), or acute leukemia.

## 2 Therapy

The current treatment algorithm is depicted in [Figure 1](#).

**Figure 1: Treatment algorithm in patients with essential thrombocythemia (ET), based on the conventional score**



**Legend:**

■ curative treatment intent; ■ non-curative treatment intent;

<sup>1</sup>Anagrelide is approved for the treatment of high-risk ET in the form of different preparations from different manufacturers, covering first- and second-line therapy in Germany, Austria and Switzerland.

<sup>2</sup>Pegylated interferon-alpha is only applicable as 'off label use'.

## 15 Authors' Affiliations

**Prof. Dr. med. Petro E. Petrides**

Hämatologisch-Onkologische Schwerpunktpraxis  
 am Isartor  
 Zweibrückenstr. 2  
 80331 München  
[petrides@onkologiemuenchen.de](mailto:petrides@onkologiemuenchen.de)

**Prof. Dr. med. Gabriela M. Baerlocher**

Universität Bern  
 Labor für Hämatopoiese und Molekulare Genetik  
 Murtenstrasse 40  
 CH-3008 Bern  
[gabriela.baerlocher@hematology.ch](mailto:gabriela.baerlocher@hematology.ch)

**Prof. Dr. med. Konstanze Döhner**

Universitätsklinikum Ulm  
Innere Medizin III  
Albert-Einstein-Allee 23  
89081 Ulm  
[konstanze.doehner@uniklinik-ulm.de](mailto:konstanze.doehner@uniklinik-ulm.de)

**Prof. Dr. med. Heinz Gisslinger**

Medizinische Universität in Wien  
Universitätsklinik f.Innere Medizin I  
Klinische Abteilung für Hämatologie und Hämostaseologie  
Währinger Gürtel 18-20  
A-1090 Wien  
[heinz.gisslinger@meduniwien.ac.at](mailto:heinz.gisslinger@meduniwien.ac.at)

**Prof. Dr. med. Martin Grieshammer**

Johannes Wesling Klinikum Minden  
Klinik für Hämatologie / Onkologie  
Hans-Nolte-Str. 1  
32429 Minden  
[martin.griesshammer@muehlenkreiskliniken.de](mailto:martin.griesshammer@muehlenkreiskliniken.de)

**Univ.-Prof. Dr. med. Steffen Koschmieder**

Uniklinik RWTH Aachen  
Med. Klinik IV  
Hämatologie, Onkologie, Hämostaseologie & SZT  
Pauwelsstr. 30  
52074 Aachen  
[skoschmieder@ukaachen.de](mailto:skoschmieder@ukaachen.de)

**PD Dr. med. Juliana Schwaab**

Universitätsmedizin Mannheim  
III. Medizinische Klinik  
Hämatologie und Internistische Onkologie  
Theodor-Kulzer-Ufer 1-3  
68167 Mannheim  
[juliana.schwaab@medma.uni-heidelberg.de](mailto:juliana.schwaab@medma.uni-heidelberg.de)

**Prof. Dr. med. Eva Lengfelder**

Universitätsklinikum Mannheim  
Medizinische Fakultät Mannheim d. Uni Heidelberg  
III. Medizinische Klinik  
Theodor-Kutzer-Ufer 1-3  
68167 Mannheim  
[eva.lengfelder@medma.uni-heidelberg.de](mailto:eva.lengfelder@medma.uni-heidelberg.de)

## **16 Disclosures**

Conflicts of interest can be found in the full [German version of the guideline](#).