

# Use and positioning of fostamatinib in the management of primary chronic immune thrombocytopenia: an Italian expert opinion

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**Abstract:** Fostamatinib, a spleen tyrosine kinase (Syk) inhibitor, represents a new therapeutic opportunity for patients with immune thrombocytopenia (ITP) in Europe and Italy. However, the positioning of this drug in patient's therapeutic sequence is undefined within the most recent international guidelines. The conclusions from a consensus meeting between Italian experts, whose task was to outline the profile of the ideal candidate to receive fostamatinib, are reported here. A modified Delphi methodology was used to achieve shared statements, which were reported in a narrative form. In particular, the panelists examined the strengths and weaknesses of the registration studies in terms of clinical outcomes, the safety profile of fostamatinib, the drug's impact on the quality of life of patients with chronic ITP, and the potential benefits of its use in the pandemic era. Although the experience with thrombopoietin receptor agonists (TPO-RAs) and the amount of data from real-world studies suggest the preferential use of these drugs as a second-line treatment in most patients, the absence of an increased thrombotic risk in the clinical trials could make fostamatinib a reasonable choice in patients with an increased risk of vascular events. An unstable platelet count during TPO-RAs might also justify a switch to the Syk inhibitor, which is more likely to stabilize the platelet count in responders. Fostamatinib may be preferred to immunosuppressors during the SARS-CoV-2 pandemic, in patients at infectious risk, or in case of contraindication to splenectomy. Finally, the novel mechanism of action makes it an attractive drug in multi-refractory patient.

**Keywords:** Fostamatinib, immune thrombocytopenia, thrombopoietin receptor agonists

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## Introduction

The most recent guidelines on the treatment of immune thrombocytopenia (ITP), in particular with respect to the management of therapy beyond 12 months [chronic ITP according to the International Working Group (IWG) criteria],<sup>1</sup> include fostamatinib among the available options,<sup>2,3</sup> but its place in the decisional chart has not yet been well defined. Fostamatinib is appropriately included in the list of drugs with robust clinical evidence according to the international consensus report.<sup>2</sup> However, the limited post-marketing experience at an international level (especially in those countries that have long waited for the commercialization and finalization

of reimbursement policies) and the current limitation of prescription, which prevents its use in the persistence phase of the disease (3–12 months) require ad hoc considerations to establish which patients can benefit most from this new therapeutic possibility. It is also essential to speculate on the correct timing of fostamatinib use within the patient-therapeutic strategy. On the contrary, the old-fashioned 'one-drug-fits-all' strategy is no longer acceptable for ITP, and we rather must consider the biological heterogeneity of the disease, the patient's comorbidities, the clinical risk, the different daily habits, and the maintenance of a sustainable quality of life as an integral part of a personalized approach strategy.<sup>4,5</sup> As a matter of

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fact, notwithstanding the high efficacy and wide use of thrombopoietin receptor agonists (TPO-RAs) in the last decade, a not negligible cohort of patients at higher thrombotic risk may benefit from alternative agents. Within the American Society of Hematology (ASH) guidelines,<sup>2</sup> patient preference finally becomes an important pillar supporting the choice of treatment. The reasoned participation of the patient and the sharing of treatment pathways with doctors is – both in Italy and in other countries of the world – the result of the lively presence of patient associations supporting the scientific community. Finally, in recent years so hard hit by the severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) pandemic, ITP experts have begun a buoyant discussion to redraw treatment strategies accounting for the infectious risk, by avoiding immunosuppression as much as possible and, in turn, to face ITP cases developing after SARS-CoV-2 infection and vaccination.<sup>6–8</sup> Here we provide a critical review of the state of the art of fostamatinib use in ITP focusing on its positioning along patient's therapy sequence and present the consideration emerged at an Italian consensus meeting.

### **Fostamatinib: drug characteristics and registration trial**

Fostamatinib is an orally administered small molecule capable of inhibiting splenic tyrosine kinase (SyK). It therefore offers a radically different mechanism of action compared to currently available therapies.<sup>9</sup> In the intestine, it is rapidly converted into its active metabolite R406. The latter inhibits FcγRs and B-cell receptor/Toll-like receptor (BCR/TLR) signal transduction, blocking the mechanisms of phagocytosis and leading to a reduction in antibody-mediated platelet (PLT) destruction. The double-blind phase 3 study against placebo<sup>10</sup> showed a stable response (Platelets  $\geq 50,000/\text{mmc}$  in at least 4 out of 6 scheduled visits every 2 weeks from Week 14 to Week 24 of the protocol) in 18% of patients. Of these, 77% had a stable platelet count at all clinical controls in the protocol. An overall response (OR, at least one platelet count  $\geq 50,000/\text{mmc}$  within 12 weeks of treatment) was achieved in 43% of patients. Response to treatment was observed within a wide range of disease history (defined as time from diagnosis of ITP to study entry). Over the 24-week evaluation period, the median count for stable responders was 95,000/

mmc, while for overall responders it was 49,000/mmc. Median time to response was 15 days, and 83% patients responded within 8 weeks, suggesting to keep on with treatment for at least 2 months. Most subjects (88%) increased their dose to 150 mg BID at or after week 4. Responses were independent of prior lines of therapy, while a higher probability of increased counts above the protocol threshold was observed in patients with circulating anti-platelet antibodies. Fostamatinib was effective in preventing bleeding events, as no bleeding-related serious adverse events (SAEs) were reported in responders (both stable and ORs), while the placebo arm was burdened by severe bleeding in 10% of patients. For the remaining SAEs, a similar rate was observed between the two arms (fostamatinib and placebo); at variance, with respect to mild and moderate events, fostamatinib was more likely to induce diarrhea, hypertension, nausea and abdominal discomfort, increased transaminases, neutropenia, and respiratory tract infections (almost exclusively mild). After the initial 24-week phase, the protocol allowed the extension of the observation in treatment or the switch to the experimental drug of patients not previously exposed (open-label extension, OLE).<sup>11</sup> This experience yielded further interesting information. The response rate remained essentially unchanged, and subjects in OLE maintained their results for more than 2 years. Data from patients who had previously failed TPO-RA are also important: 34% (24/71) of them had a recovery of their platelet count with fostamatinib. Noteworthy, concomitant medications were allowed if at a stable dose, but with an indication to taper them as far as possible. In the phase 3 study, the influence of concomitant medications was considered negligible.

### **Consensus methodology**

This article reports the results of a discussion involving eight expert panelists (EPs), invited by Grifols for participation in a consensus meeting on the basis of their clinical experience in the treatment of ITP and their research contributions on the topic. Criteria for panel qualification included practice primarily in Italy, medical specialty in hematology, >10 years practice in hematology, spending >50% of time in direct patient care and having seen a minimum number of 100 patients with ITP in the last year. The aim of the

**Table 1.** Favorable aspects and potential difficulties foreseen by the EP with respect to the introduction of fostamatinib in clinical practice.

Favorable aspects	Potential difficulties
Efficacy in multi-refractory patients Significant increase in response when employed in the second line	Available only for the chronic phase
Safe platelet counts maintained in responsive patients on most weekly controls	Possible impact of toxicity on QoL (e.g. diarrhea)
Absence of thrombosis in clinical trials; consequent possible electivity of use with respect to TPO-RA, in patients with thrombotic risk factors	Risk of neutropenia/infection/arterial hypertension
Innovative mechanism of action	'Stable' response rates quite low when administered from the third line onwards
Likely to be used in combination with other therapeutic agents, with different mechanisms of action	'Real-life' studies are lacking

EP, expert panelist; QoL, quality of life; TPO-RA, thrombopoietin receptor agonist.

meeting was to directly compare the available evidence, so as to delineate the correct positioning of fostamatinib within the framework outlined by recent international guidelines and the Italian experience in the management of ITP and possibly to profile the ideal candidate for this pharmacological treatment. During its development, the discussion on fostamatinib touched on several other topics of collective interest, such as

- a. Pivotal studies on fostamatinib: strengths, and weaknesses
- b. Safety of the drug with respect to the quality of life of the chronic ITP patient
- c. Choice of fostamatinib in the ongoing SARS-CoV-2 pandemic
- d. The need for post-marketing studies to extend the validity of efficacy and safety data outside of randomized clinical trials

Specifically, a modified Delphi methodology was employed to develop recommendations for the positioning of fostamatinib in the treatment of patients with primary ITP. Consensus statements were developed from common answers in free-text responses provided in the first-round survey:

1. What are the strengths and limitations of fostamatinib emerging from clinical trials (including efficacy and safety issue)?

2. How may the use of fostamatinib impact on quality of life of ITP patients?
3. May fostamatinib be safely used during SARS-CoV-2 pandemic?
4. Which ITP patient would you candidate to fostamatinib?

The EP produced a list of statements on fostamatinib treatment emerging from the first round of discussion. In the second-round survey, the panelists provided consensus statements reaching >80% agreement. The consensus statements are summarized thereafter in a narrative way.

Grifols did not fund nor take part to Delphi process, writing or revising the manuscript.

## Results of discussion

### *Strengths and limitations of fostamatinib use*

Table 1 highlights the 'pros' and 'cons' based on data reported in the literature on fostamatinib.

The panel noted that a haemostatically safe platelet count, conventionally >30,000/mm<sup>3</sup>, is accepted as a clinical endpoint.<sup>1</sup> The criteria defined by the registration study therefore risk penalizing the perception of the drug's real clinical usefulness. Moreover, experts also discussed the potential benefits obtained with respect to

certain treatment sequences, with particular reference to TPO-RA. This question is partially answered by the OLE study, which provides the responder rate of about 30% after failure of eltrombopag or romiplostim.

Importantly, subjects who received fostamatinib as second line (after steroids  $\pm$  immunoglobulins) in phase 3 studies were more likely to obtain a PLT response  $>50,000/\text{mmc}$  in comparison with those receiving fostamatinib as third-or-later-line therapy (78% *vs* 48%).<sup>12</sup> Counts  $\geq 30,000/\text{mmc}$  were guaranteed in 30/32 (94%) second-line patients, whereas the probability dropped to 63% (71/113) when fostamatinib was used from the third line onwards. Bleeding events were less frequent in second-line (28%) *versus* third-or-later-line (45%) patients. The probability and quality of response thus appear to be affected by early choice of drug, while response duration does not seem dependent on second- or later-line use. Although there are data on the benefits of fostamatinib in second line, some difficulties are expected in positioning the product accordingly at this stage. The main problems are related to the exclusion of the persistence phase in experimental trials and from the current indication, and to the long-standing experience with TPO-RAs, which are considered safe and effective drugs.

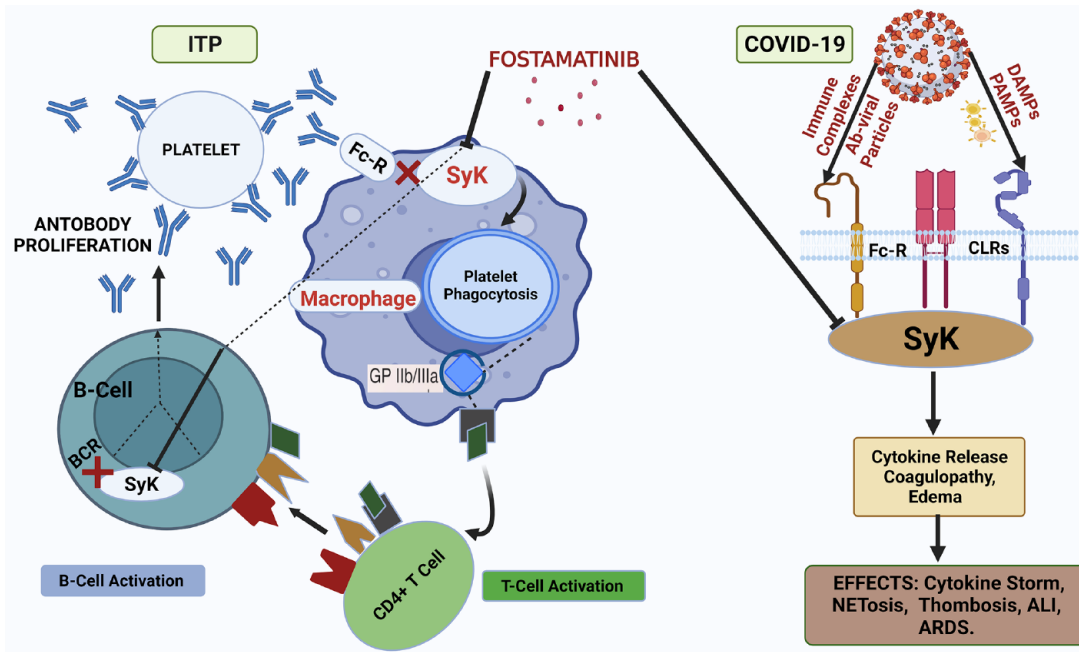
However, the panel endorses second-line use in patients with contraindications for the use of TPO-RAs and in particular in those at high thrombotic risk. In randomized studies, 146 patients received fostamatinib; 87% had at least one thrombotic risk factor and 58% more risk factors. Although patients with known thrombophilia or recent thrombosis (less than 6 months) were excluded from recruitment, some patients ( $n=11$ ) had higher risk factors such as cancer, previous thrombosis, and anti-phospholipid positivity (7.5%). A single minor episode (transient ischemic attack) was recorded in an obese and hypertensive patient, with an incidence of 0.4 per 100-year-patient on a total observation of 229-year-patient.<sup>13</sup> The concept of ITP as a 'vascular disease with increased thrombotic risk'<sup>14</sup> has been reiterated several times. In this context, it is generally believed that the use of TPO-RAs may further amplify the risk of venous and arterial thrombotic events, regardless of platelet level. In a recent meta-analysis of 740 patients with ITP enrolled in 11 randomized clinical trials and treated with TPO receptor agonists, the thrombotic risk was increased (not significantly)

by 1.82 times compared to 323 patients with ITP recruited in control arms. In eight of these studies, the history of a previous thrombosis was an exclusion criterion.<sup>15</sup> The incidence rate of thrombotic events per 100 patient-years under treatment with TPO receptor agonists resulted in 4.2 for romiplostim and 2.7 for eltrombopag.<sup>16</sup>

The thrombotic risk linked to the use of fostamatinib in clinical trials seems quite lower than that associated to the use of TPO receptor agonists, although long-term observational data would be needed for a proper comparison. The absence of thrombotic events in the aforementioned 11 very high-risk patients present in the randomized clinical trials with fostamatinib and a pilot experience in five other high-risk patients treated with fostamatinib<sup>17</sup> suggests that fostamatinib may be a treatment of choice in patients with ITP at thrombotic risk. In particular, the use of fostamatinib could be an obvious rescue treatment in patients who have suffered from thrombosis during the administration of TPO receptor agonists.

#### *Impact of fostamatinib on quality of life*

In recent years, the scientific community and patient associations have paid increasing attention to the quality of life of patients with ITP,<sup>18</sup> which is often conditioned by frequent hospital visits, limitation of activities due to the presence (or fear) of symptoms, particularly fatigue, anemia, and comorbidities, and – to a very relevant extent – also by treatments and related adverse events. In this context, fostamatinib-related adverse events such as diarrhea (occurring in 31% on fostamatinib *vs* 15% on placebo in the phase III trial), hypertension (28% *vs* 13%), nausea (19% *vs* 8%), dizziness (11% *vs* 8%), and alanine aminotransferase (ALT) increase (11% *vs* 0%) are worthy of caution. These adverse events may shadow the benefit of fostamatinib in comparison to TPO-RAs. However, data from the OLE study of fostamatinib, as well as those presented at the 2020 ASH meeting<sup>19</sup> (on over 4000 patients treated, 146 with ITP with follow-up up to 81 months), deny the appearance of new toxicities or an increase in their frequency in the long term; on the contrary, they seem to show a progressive decrease in the reporting of adverse events in the quartiles following the first year of treatment. Nevertheless, the frequency of these adverse events and possible impact on quality of life should be discussed with the patient. The panel



**Figure 1.** The downstream effects of Syk inhibition could provide benefits both in the processes of opsonisation, antigen presentation, and antibody production in ITP, and in the prevention of vascular and respiratory damage during COVID-19.

ALI, acute lung injury; ARDS, acute respiratory distress syndrome; BCR, B-cell receptor; CLR, C-type lectin receptors; DAMPs/PAMPs, disease-/pathogen-associated molecular patterns; Fc-R, Fc receptor.

suggested surveillance for mild-to-moderate elevation of blood pressure and liver enzymes that can be managed with appropriate monitoring, supportive therapy, lifestyle interventions, and dose adjustments (including those of concomitant medications).

#### *Fostamatinib use during SARS-CoV-2 pandemic*

During the consensus, the possibility that a moderate increase in the incidence of neutropenia and airway infections could have a negative impact during the COVID-19 pandemic was considered. It may be speculated that the close connection between Syk and TLR signaling (implicated in the pathogenesis of coronavirus alveolar damage)<sup>20</sup> could be an advantage in preventing the main clinical outcomes of infection (Figure 1).

Results are now available from a phase 2 study that compared fostamatinib to placebo in patients with COVID-19, hospitalized and requiring oxygen. A benefit was obtained on several clinical outcomes.<sup>21</sup> Finally, the ITP Expert Panel convened by the ASH, responding to frequently

asked questions (FAQs), recommends avoiding rituximab and immunosuppressants whenever possible, to reduce potential exposure and vulnerability, and cites fostamatinib as one of the alternative options in times of pandemic.<sup>6</sup> Moreover, great attention should be paid to the potential negative impact of immunosuppressive treatments on the seroconversion after SARS-CoV-2 vaccination. In this regard, the effect of fostamatinib is not known, and ad hoc investigation would be needed.

#### *Who may be candidate to fostamatinib?*

The designated candidate to receive fostamatinib, according to the panel discussion, may be a patient with one or more of the following characteristics:

- Need for a third-line treatment (preferably after a TPO receptor agonist) or second-line treatment in the presence of a high risk of thrombosis or other contraindications for the use of TPO-RAs.
- Contraindications to splenectomy
- Unstable counts during TPO-mimetic administration



- d. Patients in whom immunosuppression is to be avoided
- e. Refractoriness to previous lines, even with a long history of disease

Regarding the presence of anti-platelet antibodies, the panel discussed that patients carrying anti-GPIIb/IIIa antibodies, which mainly lead to Fc-independent hepatic clearance, may show lower response to fostamatinib. However, testing for anti-platelet antibodies is not formally recommended, and further evidence would be needed to support this hypothesis.

The panel considers the limitation of prescription linked to the stage of the disease (i.e. chronic phase >12 months from initial diagnosis) unduly limitative to the patient not responsive to the first line of treatment with steroids 'and' not responsive to TPO receptor agonists 'or' with high thrombotic risk. In these subjects, the time necessary to reach the indication may put the patient at high risk of disease-related (bleeding) or drug-related (thrombosis) events.

With regard to these aspects, the panel finds it appropriate to solicit a close discussion with the regulatory authorities for an extension of indication.

### Conclusion

According to the available data and the panel opinion, fostamatinib seems to offer a widely acceptable safety and handling profile. PLT increase and reduction of bleeding risk was observed in roughly 1/3 of subjects, even in challenging patients such as those exposed to multiple lines of therapy – including splenectomy and sequential immunosuppressants – and with a long history of disease. Efficacy may be possibly improved by exploring combination strategies and by the most appropriate positioning of the drug in the therapeutic algorithm. Real-world experiences will be a valuable resource for acquiring data more in line with daily practice, thus optimizing outcomes of fostamatinib-treated patients.

### Declarations

*Ethics approval and consent to participate*  
Not applicable.

### *Consent for publication*

Not applicable.

### *Author contributions*

**Alessandro Lucchesi:** Conceptualization; Data curation; Methodology; Project administration; Supervision; Writing – original draft; Writing – review & editing.

**Bruno Fattizzo:** Conceptualization; Methodology; Writing – original draft; Writing – review & editing.

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#### Availability of data and materials

This is a review article. Further information may be asked to the corresponding author.

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