

# Clinical and Hemodynamic Characterization Super Responders to IV Prostacyclin

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## Research

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# Abstract

## Background

Parenteral prostacyclins are the only therapy proven to extend survival in pulmonary arterial hypertension (PAH), yet at the bedside clinicians have no tools to predict which patients are most likely to benefit from this medication class. Methods

We retrospectively analyzed all PAH patients treated with IV epoprostenol therapy at our center from 1/1/1996 to 12/31/2016. We analyzed survival in patients and defined the 90 th percentile of survival. Patients were divided into those who survived past this point (super responders) and those who had had an event prior to this time point after initiation of iv epoprostenol (usual responders).

## Results

The median survival after IV epoprostenol initiation was 4.32 years, and the 90 th percentile of event-free survival was 11.09 years. Fourteen patients met criteria for super responder and 45 had a survival <90 th percentile, comprising the usual responder group. Super responders tended to be younger, have longer six-minute walk distances and higher mean pulmonary arterial pressure ( $p < 0.05$  for all). In follow up, super responders continued to have a higher six-minute walk distance and were more likely to have achieved normal or only mildly impaired right ventricular function, though no differences in hemodynamics were observed.

## Conclusions

There may be a super responder phenotype that can be defined in patients with PAH by >90 th percentile of survival. Super responders were more likely than usual responders to be younger and were more likely to have achieved favorable right ventricular function at follow up, however, differences in hemodynamics were not observed.

## Full Text

This preprint is available for [download as a PDF](#).

## Figures

Figure 1

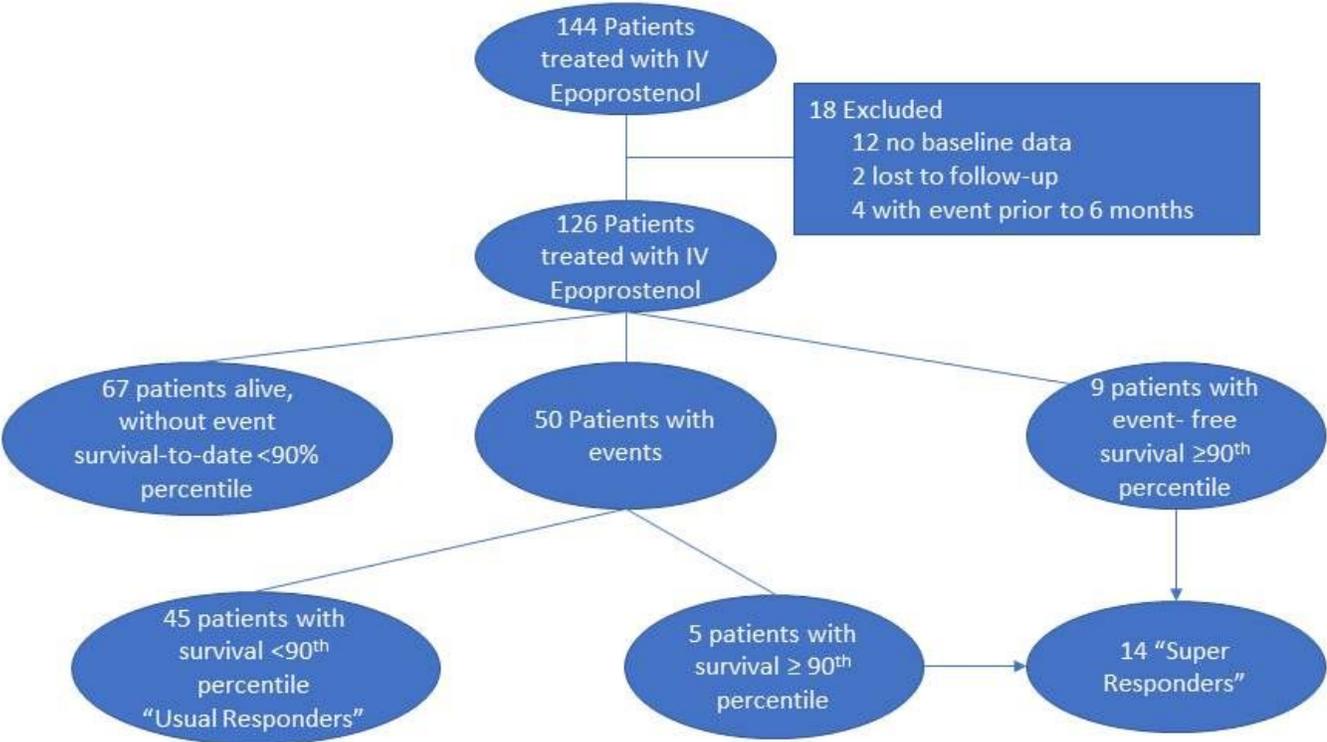


Figure 1

Patient disposition

Figure 2

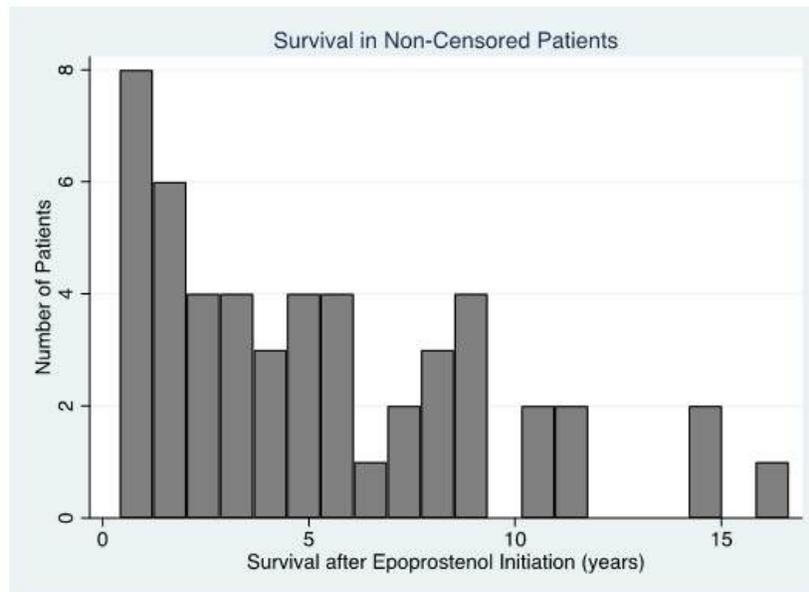


Figure 2

Survival distribution Histogram of survival in 50 patients with events in follow up showing distribution of survival.

Figure 3

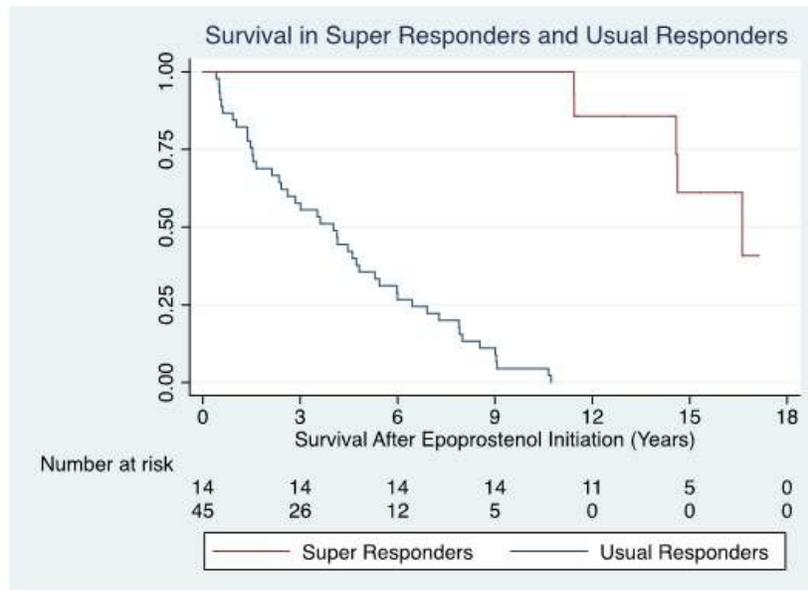


Figure 3

Survival in Super Responders and Usual Responders. The difference in survival demonstrated in super responders vs. usual responders,  $p < 0.5$ .