

## EPPVDN Abstract Rules (Symposium, Berlin 6/2022)

**Font:** Arial, 11pt

**Title:** The abstract title is limited to 200 characters without spaces. Do not use capital letters unless abbreviations or similar.

**Authors and affiliations:** Please enter authors' name and affiliations. Do not use capital letters unless abbreviations or similar.

**Grant funding and conflict of interests** should be added at the end of the abstract.

All abstracts must follow the **structure:** Background, Methods, Results and Conclusions.

Word count is limited to **250 words**. The title, authors' name and affiliation, grant funding and conflict of interests are not included in this number.

Please **do not** include the title of the abstract, the authors and affiliation, or any references in the **text**.

Please send a structured 250 word ABSTRACT in Word format **by March 31, 2022**, as Email attachment to [mail@pvdnetwork.org](mailto:mail@pvdnetwork.org). The abstract deadline may get extended by 1 month.

EXAMPLE ABSTRACT:

**Selexipag for the treatment of children with pulmonary arterial hypertension: first multicenter experience in drug safety and efficacy, on behalf of the European Pediatric Pulmonary Vascular Disease Network (EPPVDN)**

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**Background.** The European Pediatric Pulmonary Vascular Disease Network (EPPVDN) investigated the safety and efficacy of add-on selexipag, an IP prostacyclin receptor agonist approved for pulmonary arterial hypertension (PAH) in adults, in the largest pediatric cohort to date.

**Methods.** This is a prospective observational study of 15 consecutive children with PAH, treated with oral add-on selexipag at three centers. Most patients underwent cardiac catheterizations at baseline and median 8 months follow-up. All patients had clinical, echocardiographic and NTproBNP studies, including the novel EPPVDN pediatric PH risk score.

**Results.** There was no death during selexipag use. Two of 15 patients ultimately underwent lung transplantation. One patient with heritable PAH died on intravenous treprostinil (off selexipag). Mean right atrial pressure, the ratio of pulmonary artery to systemic artery pressure (mPAP/mSAP, dPAP/dSAP -17%), and transpulmonary pressure gradients (mean TPG -17%; p<0.01; diastolic TPG -6 mmHg; p<0.05) improved (n=10). Selexipag therapy was associated with better RV systolic function (TAPSE +14.5%; p<0.01) and functional class. Non-invasive and combined-invasive PH risk scores improved (lower risk + 18-22%, higher risk - 35-37%; p<0.05). Overall, the efficacy of selexipag was variable, often with better response in less sick patients.

**Conclusions.** Oral selexipag use in children with PAH is well tolerated and safe when closely monitored. Add-on selexipag therapy improved several outcome-relevant variables in about half, and prevented disease progression in additional 27% of patients. The novel EPPVDN pediatric PH risk score indicated these drug effects properly, can be useful in clinical follow-up, and should be validated in larger prospective studies.

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