Insmed Announces Positive 12-Month Data from the Ongoing Open-Label Extension Study of Treprostinil Palmitil Inhalation Powder (TPIP) in Patients with Pulmonary Arterial Hypertension

–12-Month Data Demonstrated Sustained Improvement with TPIP Across All Secondary Efficacy Measures Including Reduction in Mortality Risk Status by REVEAL Lite 2.0–

–Placebo Crossed Group Demonstrated Treatment Response on TPIP, Achieving Similar Outcomes to the TPIP Continued Group by Month 12–

–TPIP Was Safe and Well-tolerated with No Newly Identified Safety Signals through Month 12; Doses Up to 1,280 µg Once Daily Were Permitted in the OLE Study–

–These OLE Data, Combined with Once-Daily Inhaled Administration, Reinforce TPIP's Potential to Become the Prostanoid of Choice for Patients with PAH–

–Insmed to Host Investor Call Thursday, July 16, 2026,at 8:00 a.m. ET–

BRIDGEWATER, N.J., July 16, 2026 /PRNewswire/ -- Insmed Incorporated (Nasdaq: INSM), a people-first global biopharmaceutical company striving to deliver first- and best-in-class therapies to transform the lives of patients facing serious diseases, today announced positive 12-Month data from the ongoing open-label extension (OLE) study evaluating treprostinil palmitil inhalation powder (TPIP), administered once daily in patients with pulmonary arterial hypertension (PAH, World Health Organization Group 1). The OLE study is a non-placebo-controlled trial and was designed to evaluate the long-term safety, tolerability, and effectiveness of TPIP over 24 months in patients who completed the lead-in TPIP PAH studies.

"These data from our ongoing OLE study with TPIP represent an important milestone in our efforts to fully harness the potential of treprostinil and provide meaningful benefit to patients with pulmonary arterial hypertension," said Gene Sullivan, M.D., Chief Product Strategy Officer of Insmed. "In this analysis, TPIP demonstrated sustained improvement across all efficacy endpoints and was well tolerated with no newly identified safety signals, and notably, patients who switched from placebo to TPIP in the OLE study achieved similar clinical benefit. These data, coupled with the statistically significant and clinically meaningful results from our Phase 2b randomized study, demonstrate TPIP's potential to become the prostanoid of choice for PAH patients, and we remain excited to be advancing our Phase 3 PALM-PAH study."

12-Month Results from the Ongoing OLE Study with TPIP in Patients with PAH
Results for secondary efficacy endpoints demonstrated sustained improvement with TPIP in six-minute walk distance (6MWD), N-terminal fragment pro-B-type natriuretic peptide (NT-proBNP) concentration, and World Health Organization (WHO) Functional Class, as well as a clinically meaningful improvement in REVEAL Lite 2.0 score at Month 12. Patients in the Placebo Crossed group (N=31) showed similar outcomes to patients in the TPIP Continued group (N=60) across all efficacy measures at Month 12.

The results at Month 12 were as follows:

- Mean improvement from baseline for 6MWD was +55.7 meters for the TPIP Continued group and +54.1 meters for the Placebo Crossed group.
- NT-proBNP concentration was reduced by approximately 60% in both groups with geometric mean ratios [GMR] to baseline of 0.40 and 0.41 in TPIP Continued and Placebo Crossed, respectively.
- WHO Functional Class I or II was achieved in 78.3% of TPIP Continued group and 80.6% of Placebo Crossed group patients, and more than 25% of patients across both groups achieved WHO Functional Class I.
- Mean REVEAL Lite 2.0 score improved 2.0-points from baseline for the TPIP Continued group and 1.4-points from baseline for the Placebo Crossed group. Approximately 65% of all patients achieved Refined Low Risk status, which is associated with a less than 5% estimated risk of mortality at three years and an approximately 7% risk of clinical

worsening at one year.

"Pulmonary arterial hypertension is one of the most devastating diseases we face as clinicians, and these results from the ongoing TPIP OLE study give us genuine reason for optimism," said Dr. Raymond Benza, M.D., F.A.C.C., FAHA, FACP, Phase 2b PAH study Steering Committee Member, George M. and Linda H. Kaufman Academic Chair of Cardiology, Sentara Health. "The REVEAL Lite 2.0 risk score provides a powerful, non-invasive way to track disease trajectory. Previous REVEAL Lite 2.0 validation showed that a 1-point score improvement reduces risk of mortality by 23% and reduces the risk of clinical worsening by 21%. Here we saw that patients on TPIP averaged a greater than 1-point improvement from baseline, which is really meaningful for patients. Sustained improvements of this magnitude, alongside gains in exercise capacity and Functional Class, provide a strong clinical rationale to advance this therapy into Phase 3 development."

Results for the primary endpoint of safety & tolerability showed that once-daily TPIP therapy was generally well tolerated with no newly identified safety signals at doses up to 1,280 µg through Month 12. Of the 91 patients in the study, treatment-emergent adverse events (TEAEs) occurred in 89.0% of patients; serious TEAEs were observed in 18.7% of patients; and severe TEAEs were observed in 16.5% of patients in the study. TEAEs leading to study discontinuation were experienced by 7.7% of patients. There were four deaths, none of which were considered related to TPIP treatment. The most common TEAEs through Month 12 occurring in 5.0% or more of all patients were headache (28.6%), cough (15.4%), nasopharyngitis (14.3%), diarrhea (11.0%), upper respiratory tract infection (9.9%), bronchitis (7.7%), dizziness (6.6%), epistaxis (6.6%), nausea (6.6%), anemia (5.5%), influenza (5.5%), and pneumonia (5.5%).

The 12-Month OLE findings support the continued clinical development of TPIP and the recent initiation of PALM-PAH, a Phase 3, randomized, double-blind, placebo-controlled trial evaluating once-daily TPIP in patients with PAH over 24 weeks. The primary endpoint of PALM-PAH is change in 6MWD, with additional assessments of safety, tolerability, and overall efficacy. Insmed plans to publish the 12-Month results from this OLE study in the future. Topline results from the Phase 2b study of TPIP in patients with PAH were previously reported in June 2025.

Conference Call Information

Insmed management will host a conference call for investors beginning at 8:00 a.m. ET on Thursday, July 16, 2026, to discuss the TPIP OLE study results. Shareholders and other interested parties may participate in the conference call by dialing (800) 715-9871 (U.S.) or +1 (646) 307-1963 (International) and referencing access code 2033335. The call will also be webcast live on the Company's website at www.insmed.com.

A replay of the conference call will be accessible approximately one hour after its completion through July 23, 2026, by dialing (800) 770-2030 (U.S. and Canada) or +1 (609) 800-9909 (International) and referencing access code 2033335. A webcast of the call will also be archived for 90 days under the Investor Relations section of the Company's website at www.insmed.com.

About TPIP

Treprostinil palmitil inhalation powder (TPIP) is a dry powder formulation of treprostinil palmitil, a treprostinil prodrug consisting of treprostinil linked by an ester bond to a 16-carbon chain. Developed entirely in Insmed's laboratories, TPIP is a potentially highly differentiated prostanoid being evaluated as a once-daily therapy for the treatment of patients with PAH, pulmonary hypertension associated with interstitial lung disease (PH-ILD), progressive pulmonary fibrosis (PPF), and idiopathic pulmonary fibrosis (IPF). TPIP is administered in a capsule-based inhalation device. TPIP is an investigational drug product that has not been approved for any indication in any jurisdiction.

About the TPIP Open-Label Extension Study

The 24-month open-label extension (OLE) study of treprostinil palmitil inhalation powder (TPIP) in patients with pulmonary arterial hypertension (PAH) was designed to evaluate the long-term safety, tolerability, and effectiveness of TPIP administered once daily in patients who completed the lead-in TPIP PAH studies. The OLE is being conducted at 45 sites globally and enrolled 91 eligible patients. The study included a 3-week blinded titration period followed by open-label treatment. Patients who received TPIP in the lead-in studies (TPIP Continued) received their achieved dose of TPIP; patients who received placebo in the lead-in studies (Placebo Crossed), or had delayed rollover, underwent titration starting at 80 µg once daily up to their target dose (i.e., 640 µg or highest tolerated dose) over three weeks. Further escalation up to 1,280 µg was permitted after the initial titration period at investigator discretion to optimize clinical benefit. Outcomes were evaluated against the Phase 2b lead-in study in PAH (NCT05147805) pre-randomization baseline values for relevant measurements.

The primary endpoint is evaluating long-term safety and tolerability, including treatment-emergent adverse events (TEAEs) and TEAEs by severity. Secondary endpoints include change from lead-in study pre-randomization baseline in six-minute walk distance (6MWD), N-terminal fragment pro-B-type natriuretic peptide (NT-proBNP) concentration, World Health Organization (WHO) Functional Class, REVEAL Lite 2.0 score, and additional exploratory measures over 24 months of treatment.

About Pulmonary Arterial Hypertension

Pulmonary arterial hypertension (PAH) is a serious, progressive, rare disease in which the blood vessels in the lungs narrow or

become obstructed, leading to high blood pressure in the pulmonary arteries. The most common symptoms include shortness of breath, chest pain, dizziness or fainting, fatigue, and weakness. It is estimated that approximately 35,000 patients in the U.S., 40,000 patients in the EU_5 (France, Germany, Italy, Spain, and the UK), and 15,000 patients in Japan have been diagnosed with the disease. Untreated PAH can be debilitating and often fatal.

About Insmed

Insmed Incorporated is a people-first global biopharmaceutical company striving to deliver first- and best-in-class therapies to transform the lives of patients facing serious diseases. The Company is advancing a diverse portfolio of approved and mid- to late-stage investigational medicines—including two approved therapies to treat chronic, debilitating lung diseases—as well as cutting-edge drug discovery focused on serving patient communities where the need is greatest. Insmed's commercial portfolio and clinical pipeline are organized around three therapeutic areas: Respiratory, Immunology & Inflammation, and Neuro & Other Rare. The Company's research engine is advancing a wide range of technologies and modalities, including gene therapy, AI-driven protein engineering, RNA end-joining, and synthetic rescue, in the pursuit of future pipeline candidates.

Headquartered in Bridgewater, New Jersey, Insmed has offices and research locations throughout the United States, Europe, and Japan. Insmed is proud to be recognized as one of the best employers in the biopharmaceutical industry, including spending five consecutive years as the No. 1 Science Top Employer. Visit **www.insmed.com** to learn more or follow us on **LinkedIn**, **Instagram**, **YouTube**, and **X**.

Forward-looking Statements

This press release contains forward-looking statements that involve substantial risks and uncertainties. "Forward-looking statements," as that term is defined in the Private Securities Litigation Reform Act of 1995, are statements that are not historical facts and involve a number of risks and uncertainties. Words herein such as "may," "will," "should," "could," "would," "expects," "plans," "anticipates," "believes," "estimates," "projects," "predicts," "intends," "potential," "continues," and similar expressions (as well as other words or expressions referencing future events, conditions or circumstances) may identify forward-looking statements.

The forward-looking statements in this press release are based upon the Company's current expectations and beliefs, and involve known and unknown risks, uncertainties and other factors, which may cause the Company's actual results, performance and achievements and the timing of certain events to differ materially from the results, performance, achievements or timings discussed, projected, anticipated or indicated in any forward-looking statements. Such risks, uncertainties and other factors include, among others, the following: the risk that the full data set from the OLE study of TPIP in PAH (the "Study") or data generated in further clinical trials of TPIP will not be consistent with the interim results of the Study; the risk that data from the Study, which is conducted in a single-arm, open-label design without a concurrent placebo control group, may not be predictive of, or may differ materially from, results obtained in the Phase 3 PALM-PAH randomized, double-blind, placebo-controlled trial; the risk that the Study's efficacy endpoints, which are secondary and exploratory in nature, may overestimate or otherwise not accurately reflect the true treatment effect of TPIP; failure to successfully conduct future clinical trials for TPIP, such as the Company's planned Phase 3 program for TPIP, including due to the Company's potential inability to enroll or retain sufficient patients to conduct and complete the trials or generate data necessary for regulatory approval, among other things; development of unexpected safety or efficacy concerns related to TPIP; failure of third parties on which the Company is dependent to manufacture sufficient quantities of TPIP for clinical needs, to conduct the Company's clinical trials, or to comply with the Company's agreements or laws and regulations that impact the Company's business or agreements with the Company; failure to obtain regulatory approval for TPIP; inaccuracies in the Company's estimates of the size of the potential markets for TPIP or in data the Company has used to identify physicians; expected rates of patient uptake, duration of expected treatment, or expected patient adherence or discontinuation rates, if TPIP is approved; inability of the Company or the Company's third-party manufacturers to comply with regulatory requirements related to TPIP; the Company's inability to obtain adequate reimbursement from government or third-party payors for TPIP or acceptable prices for TPIP, if approved; restrictions or other obligations imposed on the Company by agreements related to TPIP and failure to comply with the Company's obligations under such agreements; risks that the Company's clinical studies will be delayed or that serious side effects will be identified during drug development; the strength and enforceability of the Company's intellectual property rights or the rights of third parties; and the cost and potential reputational damage resulting from litigation to which the Company may become a party, including product liability claims.

The Company may not actually achieve the results, plans, intentions or expectations indicated by the Company's forward-looking statements because, by their nature, forward-looking statements involve risks and uncertainties because they relate to events and depend on circumstances that may or may not occur in the future. For additional information about the risks and uncertainties that may affect the Company's business, please see the factors discussed in Item 1A, "Risk Factors," in the Company's Annual Report on Form 10-K for the year ended December 31, 2025 and any subsequent Company filings with the Securities and Exchange Commission (SEC).

The Company cautions readers not to place undue reliance on any such forward-looking statements, which speak only as of the date of this press release. The Company disclaims any obligation, except as specifically required by law and the rules of the SEC, to publicly update or revise any such statements to reflect any change in expectations or in events, conditions or

circumstances on which any such statements may be based, or that may affect the likelihood that actual results will differ from those set forth in the forward-looking statements.

Contact:

Investors:
Bryan Dunn
Vice President, Investor Relations
(646) 812-4030
investor.relations@insmed.com

Media:
Claire Mulhearn
Vice President, Corporate Communications
(862) 842-6819
media@insmed.com

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