Insmed Reports First-Quarter 2024 Financial Results and Provides Business Update

— ARIKAYCE® (amikacin liposome inhalation suspension) Total Revenue of $75.5 Million for the First Quarter of 2024, Reflecting 16% Annual Growth Over the First Quarter of 2023—

—Company Reports Positive Topline Safety and Tolerability Data from the Phase 2 PH-ILD Study of TPIP with 79.3% of Patients Reaching the Maximum Dose of 640 µg by Week 5, with an Unexpectedly Positive and Robust Signal on the Exploratory Endpoint of Clinical Worsening—

—Encouraging Blinded Data from First 40 Patients in Phase 2 PAH Study of TPIP, with Combined Active Treatment and Placebo Arms Showing Mean PVR Reduction of 19.9% and 6-Minute Walk Distance Improvement of 43 Meters—

—Topline Data from the Phase 3 ASPEN Trial of Brensocatib in Patients with Bronchiectasis Remains on Track to Read Out in the Latter Part of Second-Quarter 2024—

—Company Reiterates 2024 Global ARIKAYCE Revenue Guidance in the Range of $340 Million to $360 Million, Reflecting Double-Digit Growth Compared to 2023—

BRIDGEWATER, N.J., May 9, 2024 /PRNewswire/ -- Insmed Incorporated (Nasdaq:INSM), a global biopharmaceutical company on a mission to transform the lives of patients with serious and rare diseases, today reported financial results for the first quarter ended March 31, 2024 and provided a business update.

"Today, Insmed kicked off a series of crucial data readouts, starting with TPIP, where we continue to observe a favorable safety and tolerability profile from the Phase 2 PH-ILD trial results as well as encouraging additional blinded PVR data from the Phase 2 PAH trial. Together, we believe these results continue to position TPIP as a potential best-in-class prostanoid," said Will Lewis, Chair and Chief Executive Officer of Insmed. "We now await with great anticipation the ASPEN trial results, which are expected in the latter half of this quarter. Supporting our research efforts is the ARIKAYCE commercial engine, which produced double-digit revenue growth this quarter, keeping us on track to meet our 2024 revenue guidance."

Recent Pillar Highlights

Pillar 1: ARIKAYCE

- ARIKAYCE global revenue grew 16% in the first quarter of 2024 compared to the first quarter of 2023, reflecting double-digit year-over-year growth in the U.S., Japan, and Europe.
- The Company received notification that the National Agency for the Safety of Medicines and Health in France plans to proceed with a program to allow for the compassionate prescribing of ARIKAYCE for the treatment of adult patients with *mycobacterium abscessus* lung infection.
- The Company is scheduled to meet with the U.S. Food and Drug Administration (FDA) in late June to gain additional insights and guidance on the patient-reported outcome tool to be used in the Phase 3 ENCORE study, after which it will finalize its statistical plan for ENCORE.
- The Data Safety Monitoring Committee for the ongoing ENCORE trial held its fourth safety review meeting in April of 2024 and recommended that the study progress unchanged.
- The Company continues to expect topline data from ENCORE in 2025.

Pillar 2: Brensocatib

- Insmed continues to expect topline data from the Phase 3 ASPEN study of brensocatib in patients with non-cystic fibrosis bronchiectasis (NCFBE) in the latter part of the second quarter of 2024.
- As of the end of the first quarter of 2024, all adult patients in ASPEN had completed their 52-week visit, the point in the trial at which the primary and secondary efficacy endpoints are measured.
- If ASPEN is successful and regulatory approval is obtained, the Company anticipates a launch in the U.S. in mid-2025, followed by launches in Europe and Japan in the first half of 2026. Insmed continues to advance its launch readiness

activities in preparation for these potential launches.
- In the first quarter of 2024, Insmed received notification from the Medicines and Healthcare products Regulatory Agency in the United Kingdom regarding a positive outcome for the Company's Innovative Licensing and Access Pathway (ILAP) passport application, granting Innovation Passport designation for brensocatib in the treatment of NCFBE in patients aged 12 years and older. The ILAP and the Innovation Passport are intended to accelerate time to market, facilitating patient access for important new medicines in the United Kingdom.
- The Company continues to enroll patients in the Phase 2b BiRCh trial of brensocatib in patients with chronic rhinosinusitis without nasal polyps (CRSsNP) and anticipates providing topline data from the study in 2025.
- The Company expects to initiate a Phase 2 study of brensocatib in patients with hidradenitis suppurativa (HS) in the second half of 2024, pending positive results from the ASPEN study.

Pillar 3: TPIP

PH-ILD

- Today, Insmed reported topline safety and tolerability data as well as certain exploratory efficacy endpoints from the Phase 2 study of treprostinil palmitil inhalation powder (TPIP) in patients with pulmonary hypertension associated with interstitial lung disease (PH-ILD).
    - In the TPIP arm, 79.3% of patients were successfully able to reach the maximum 640 microgram (µg) dose by Week 5, compared to 100.0% of patients in the placebo arm.
    - Treatment-emergent adverse events which led to treatment discontinuation occurred in 13.8% of patients in the active treatment arm and 30.0% of patients in the placebo arm.
    - Adverse events of any kind were observed in 93.1% of patients in the TPIP arm and 90.0% of patients in the placebo arm. Adverse events related to study drug occurred in 55.2% of TPIP patients and 40.0% of placebo patients.
        - Study drug-related cough was observed in 37.9% of patients in the TPIP arm and 0.0% of patients in the placebo group.
        - All events of cough were mild, and none led to treatment discontinuation.
    - Serious adverse events occurred in 20.7% of TPIP-treated patients and 40.0% of placebo-treated patients.
    - Deaths occurred in 6.9% of patients taking TPIP and 20.0% of patients taking placebo.
        - All deaths were attributed to disease progression or comorbid causes, none of which were deemed related to study drug.
    - There were no meaningful changes in oxygenation levels compared to baseline for TPIP-treated patients at rest or at the lowest point during or after exercise. There was also no change in the use of supplemental oxygen for patients taking TPIP.
        - There was a small decrease in oxygenation levels observed after exercise for patients on TPIP, compared to a slight increase for patients taking placebo. However, there was variability in the timing of when this endpoint was measured in the study, which could make the results less interpretable than the measurements taken at rest or at the lowest point during or after exercise.
    - On the exploratory endpoint of change from baseline in 6-minute walk distance, TPIP-treated patients demonstrated a 30-meter improvement compared to patients treated with placebo. However, this result was associated with a wide confidence interval.
    - There was a directional improvement observed in NT-proBNP levels from baseline for patients taking TPIP and a directional worsening observed in patients on placebo, although no meaningful separation was observed between groups.
    - Events of clinical worsening occurred in 10.3% of patients taking TPIP, compared to 50.0% of patients taking placebo. This difference was nominally significant (p=0.0164).
- Due to the small size of the study, the Company interprets these results with appropriate caution.
- Insmed looks forward to the opportunity to present pharmacokinetic results and additional safety and exploratory endpoints from this trial at an upcoming medical conference later this year.
- Based on these Phase 2 results in PH-ILD, the Company is advancing toward discussions with global regulatory authorities on the design of a Phase 3 study in PH-ILD, which the Company anticipates initiating in 2025.

PAH

- Today, the Company also shared updated blinded and blended data from the first 40 patients who completed the full 16 weeks of treatment in the ongoing Phase 2 study of TPIP in pulmonary arterial hypertension (PAH).

- Of those 40 patients, including those on placebo, the average reduction in pulmonary vascular resistance (PVR) at Week 16 compared to baseline levels was 19.9%.
- Among the 40 patients across the treatment and placebo arms, the average improvement in 6-minute walk distance from baseline was 43 meters.
- Among the first 43 patients to complete the Week 5 visit, 79.1% were able to reach the maximum dose of 640 µg or matching placebo.

- Insmed has received the necessary regulatory approvals in 10 of 17 countries where the study is taking place to amend the protocol to allow for an increase in the maximum dose of TPIP from 640 µg to up to 1,280 µg, once a day, in the open label extension study for certain PAH patients based on investigator decision.
- Enrollment remains ongoing in the Phase 2 PAH study, with more than half of the target enrollment currently complete.
- In March of 2024, the second meeting of the Data Monitoring Committee took place for the Phase 2 trial in PAH, resulting in the committee's recommendation to continue the trial without changes.
- Topline results from the Phase 2 PAH study continue to be expected in 2025.

Pillar 4: Early-Stage Research

- Insmed's early-stage research efforts include more than 30 identified pre-clinical programs in development, all of which have the potential to become first-in-class or best-in-class therapies.
- The Company continues to anticipate the totality of its early-stage research programs will comprise less than 20% of overall spend.

Corporate Updates

- Insmed plans to present nine abstracts from across its respiratory portfolio (ARIKAYCE, brensocatib, and TPIP) at the American Thoracic Society (ATS) 2024 International Conference, taking place May 17-22, 2024.
- The Company served as the founding sponsor of the COPD Foundation's new Care Center Network for patients with bronchiectasis and nontuberculous mycobacterial lung disease. Through this initiative, the COPD Foundation aims to create 150 multi-disciplinary centers of excellence across the U.S. to establish consistent standards of care coming from expert-led academic centers and share them with the broader community in an effort to bring more comprehensive care to patients as they strive to meet treatment goals.
- Insmed's Chief Medical Officer, Martina Flammer, M.D., M.B.A., has been appointed Chair of the New Jersey Rare Disease Advisory Council (RDAC) by Governor Phil Murphy. In an effort established by the National Organization for Rare Disorders (NORD), RDACs bring together a unified, multidisciplinary network of state-wide experts focused on raising awareness of rare diseases and funding much-needed new research. Martina was nominated by BioNJ, the life sciences trade association for New Jersey, to represent the biopharma industry in New Jersey's RDAC.

First-Quarter 2024 Financial Results

- Total revenue for the first quarter ended March 31, 2024 was $75.5 million, reflecting 16% growth compared to total revenue of $65.2 million for the first quarter of 2023.
- Total revenue for first-quarter 2024 was comprised of ARIKAYCE net sales of $56.3 million in the U.S., $14.9 million in Japan, and $4.3 million in Europe and rest of world. First-quarter 2024 sales demonstrated year-over-year growth of 15% in the U.S., 13% in Japan, and 42% in Europe and rest of world, reflecting continued growth trends for ARIKAYCE in these regions.
- Cost of product revenues (excluding amortization of intangibles) was $17.5 million for the first quarter of 2024, compared to $13.8 million for the first quarter of 2023, primarily reflecting increased sales volumes of ARIKAYCE.
- Research and development (R&D) expenses were $121.1 million for the first quarter of 2024, compared to $127.9 million for the first quarter of 2023, a decrease that reflects a non-cash charge of $10.3 million related to the acquisition of Vertuis Bio, Inc. in the first quarter of 2023.
- Selling, general and administrative (SG&A) expenses for the first quarter of 2024 were $93.1 million, compared to $79.9 million for the first quarter of 2023. The year-over-year increase in SG&A expenses resulted primarily from increases in compensation and benefit-related expenses.
- For the first-quarter 2024, Insmed reported a net loss of $157.1 million, or $1.06 per share, compared to a net loss of $159.8 million, or $1.17 per share, for the first-quarter 2023.

Balance Sheet, Financial Guidance, and Planned Investments

- As of March 31, 2024, Insmed had cash and cash equivalents totaling $595.7 million.
- Insmed is reiterating its sales guidance for full-year 2024 global ARIKAYCE revenues in the range of $340 million to $360 million, representing 15% year-over-year growth at the midpoint compared to 2023.
- Insmed continues to anticipate that over 80% of total expenditures will be on its mid- to late-stage and commercial programs (ARIKAYCE, brensocatib, and TPIP), and that less than 20% of overall spend will be on its early-stage research programs, reflecting the Company's historical approach to spending.
- The Company plans to continue to invest in the following key activities in 2024:

(i) commercialization and expansion of ARIKAYCE globally;
(ii) advancement of brensocatib, including the Phase 3 ASPEN study in patients with bronchiectasis, commercial launch readiness activities, the ongoing Phase 2 trial in patients with CRSsNP, and the Phase 2 program in HS to be initiated in the second half of the year if the ASPEN result is positive;
(iii) advancement of the clinical trial program for ARIKAYCE, which is intended to satisfy the post-marketing requirement for full approval of its current indication and potentially support label expansion to include all patients with a MAC lung infection;
(iv) advancement of its clinical development programs for TPIP; and
(v) development of its early-stage research programs.

Conference Call

Insmed will host a conference call beginning today at 8:00 AM Eastern Time. Shareholders and other interested parties may participate in the conference call by dialing (888) 210-2654 (U.S.) and (646) 960-0278 (international) and referencing access code 7862189. 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 1 hour after its completion through June 8, 2024, by dialing (800) 770-2030 (U.S.) and (609) 800-9909 (international) and referencing access code 7862189. 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**.

INSMED INCORPORATED
Consolidated Statements of Net Loss
(in thousands, except per share data)
(unaudited)

	Three Months Ended March 31,	
	2024	**2023**
Product revenues, net	$ 75,500	$ 65,214
Operating expenses:		
Cost of product revenues (excluding amortization of intangible assets)	17,457	13,830
Research and development	121,083	127,865
Selling, general and administrative	93,102	79,914
Amortization of intangible assets	1,263	1,263
Change in fair value of deferred and contingent consideration liabilities	(11,900)	(9,500)
Total operating expenses	221,005	213,372
Operating loss	(145,505)	(148,158)
Investment income	8,783	10,524
Interest expense	(21,042)	(20,003)
Change in fair value of interest rate swap	2,362	(1,533)
Other expense, net	(1,100)	(111)
Loss before income taxes	(156,502)	(159,281)
Provision for income taxes	589	483
Net loss	$ (157,091)	$ (159,764)
Basic and diluted net loss per share	$ (1.06)	$ (1.17)
Weighted average basic and diluted common shares outstanding	148,456	136,355

INSMED INCORPORATED
Consolidated Balance Sheets
(in thousands, except par value and share data)

	As of March 31, 2024 (unaudited)	As of December 31, 2023
Assets		
Current assets:		
Cash and cash equivalents	$ 595,729	$ 482,374
Marketable securities	-	298,073
Accounts receivable	37,162	41,189
Inventory	82,957	83,248
Prepaid expenses and other current assets	42,874	24,179
Total current assets	758,722	929,063
Fixed assets, net	68,660	65,384
Finance lease right-of-use assets	20,307	20,985
Operating lease right-of-use assets	17,157	18,017
Intangibles, net	62,441	63,704
Goodwill	136,110	136,110
Other assets	95,698	96,574
Total assets	$ 1,159,095	$ 1,329,837
Liabilities and shareholders' equity		
Current liabilities:		
Accounts payable and accrued liabilities	$ 189,362	$ 214,987
Current portion of long-term debt	224,194	-
Finance lease liabilities	2,695	2,610
Operating lease liabilities	4,609	8,032
Total current liabilities	420,860	225,629
Debt, long-term	939,081	1,155,313
Royalty financing agreement	156,967	155,034
Contingent consideration	63,700	84,600
Finance lease liabilities, long-term	26,320	27,026
Operating lease liabilities, long-term	13,809	11,013
Other long-term liabilities	3,166	3,145
Total liabilities	1,623,903	1,661,760
Shareholders' equity:		
Common stock, $0.01 par value; 500,000,000 authorized shares, 148,560,882 and 147,977,960 issued and outstanding shares at March 31, 2024 and December 31, 2023, respectively	1,486	1,480
Additional paid-in capital	3,138,578	3,113,487
Accumulated deficit	(3,603,236)	(3,446,145)
Accumulated other comprehensive loss	(1,636)	(745)
Total shareholders' deficit	(464,808)	(331,923)
Total liabilities and shareholders' deficit	$ 1,159,095	$ 1,329,837

About ARIKAYCE

ARIKAYCE is approved in the United States as ARIKAYCE® (amikacin liposome inhalation suspension), in Europe as ARIKAYCE® Liposomal 590 mg Nebuliser Dispersion, and in Japan as ARIKAYCE® inhalation 590 mg (amikacin sulfate inhalation drug product). Current international treatment guidelines recommend the use of ARIKAYCE for appropriate patients. ARIKAYCE is a novel, inhaled, once-daily formulation of amikacin, an established antibiotic that was historically administered intravenously and associated with severe toxicity to hearing, balance, and kidney function. Insmed's proprietary PULMOVANCE® liposomal technology enables the delivery of amikacin directly to the lungs, where liposomal amikacin is taken

up by lung macrophages where the infection resides, while limiting systemic exposure. ARIKAYCE is administered once daily using the Lamira® Nebulizer System manufactured by PARI Pharma GmbH (PARI).

About PARI Pharma and the Lamira® Nebulizer System

ARIKAYCE is delivered by a novel inhalation device, the Lamira® Nebulizer System, developed by PARI. Lamira® is a quiet, portable nebulizer that enables efficient aerosolization of ARIKAYCE via a vibrating, perforated membrane. Based on PARI's 100-year history working with aerosols, PARI is dedicated to advancing inhalation therapies by developing innovative delivery platforms to improve patient care.

About Brensocatib

Brensocatib is a small molecule, oral, reversible inhibitor of dipeptidyl peptidase 1 (DPP1) being developed by Insmed for the treatment of patients with bronchiectasis, CRSsNP, and other neutrophil-mediated diseases. DPP1 is an enzyme responsible for activating neutrophil serine proteases (NSPs), such as neutrophil elastase, in neutrophils when they are formed in the bone marrow. Neutrophils are the most common type of white blood cell and play an essential role in pathogen destruction and inflammatory mediation. In chronic inflammatory lung diseases, neutrophils accumulate in the airways and result in excessive active NSPs that cause lung destruction and inflammation. Brensocatib may decrease the damaging effects of inflammatory diseases such as bronchiectasis by inhibiting DPP1 and its activation of NSPs. Brensocatib is an investigational drug product that has not been approved for any indication in any jurisdiction.

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 for the treatment of patients with PAH, PH-ILD, and other rare and serious pulmonary disorders. 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.

IMPORTANT SAFETY INFORMATION AND BOXED WARNING FOR ARIKAYCE IN THE U.S.

WARNING: RISK OF INCREASED RESPIRATORY ADVERSE REACTIONS

ARIKAYCE has been associated with an increased risk of respiratory adverse reactions, including hypersensitivity pneumonitis, hemoptysis, bronchospasm, and exacerbation of underlying pulmonary disease that have led to hospitalizations in some cases.

Hypersensitivity Pneumonitis has been reported with the use of ARIKAYCE in the clinical trials. Hypersensitivity pneumonitis (reported as allergic alveolitis, pneumonitis, interstitial lung disease, allergic reaction to ARIKAYCE) was reported at a higher frequency in patients treated with ARIKAYCE plus background regimen (3.1%) compared to patients treated with a background regimen alone (0%). Most patients with hypersensitivity pneumonitis discontinued treatment with ARIKAYCE and received treatment with corticosteroids. If hypersensitivity pneumonitis occurs, discontinue ARIKAYCE and manage patients as medically appropriate.

Hemoptysis has been reported with the use of ARIKAYCE in the clinical trials. Hemoptysis was reported at a higher frequency in patients treated with ARIKAYCE plus background regimen (17.9%) compared to patients treated with a background regimen alone (12.5%). If hemoptysis occurs, manage patients as medically appropriate.

Bronchospasm has been reported with the use of ARIKAYCE in the clinical trials. Bronchospasm (reported as asthma, bronchial hyperreactivity, bronchospasm, dyspnea, dyspnea exertional, prolonged expiration, throat tightness, wheezing) was reported at a higher frequency in patients treated with ARIKAYCE plus background regimen (28.7%) compared to patients treated with a background regimen alone (10.7%). If bronchospasm occurs during the use of ARIKAYCE, treat patients as medically appropriate.

Exacerbations of underlying pulmonary disease has been reported with the use of ARIKAYCE in the clinical trials. Exacerbations of underlying pulmonary disease (reported as chronic obstructive pulmonary disease (COPD), infective exacerbation of COPD, infective exacerbation of bronchiectasis) have been reported at a higher frequency in patients treated with ARIKAYCE plus background regimen (14.8%) compared to patients treated with background regimen alone (9.8%). If exacerbations of underlying pulmonary disease occur during the use of ARIKAYCE, treat patients as medically appropriate.

Anaphylaxis and Hypersensitivity Reactions: Serious and potentially life-threatening hypersensitivity reactions, including anaphylaxis, have been reported in patients taking ARIKAYCE. Signs and symptoms include acute onset of skin and mucosal tissue hypersensitivity reactions (hives, itching, flushing, swollen lips/tongue/uvula), respiratory difficulty (shortness of breath, wheezing, stridor, cough), gastrointestinal symptoms (nausea, vomiting, diarrhea, crampy abdominal pain), and cardiovascular signs and symptoms of anaphylaxis (tachycardia, low blood pressure, syncope, incontinence, dizziness). Before therapy with

ARIKAYCE is instituted, evaluate for previous hypersensitivity reactions to aminoglycosides. If anaphylaxis or a hypersensitivity reaction occurs, discontinue ARIKAYCE and institute appropriate supportive measures.

Ototoxicity has been reported with the use of ARIKAYCE in the clinical trials. Ototoxicity (including deafness, dizziness, presyncope, tinnitus, and vertigo) were reported with a higher frequency in patients treated with ARIKAYCE plus background regimen (17%) compared to patients treated with background regimen alone (9.8%). This was primarily driven by tinnitus (7.6% in ARIKAYCE plus background regimen vs 0.9% in the background regimen alone arm) and dizziness (6.3% in ARIKAYCE plus background regimen vs 2.7% in the background regimen alone arm). Closely monitor patients with known or suspected auditory or vestibular dysfunction during treatment with ARIKAYCE. If ototoxicity occurs, manage patients as medically appropriate, including potentially discontinuing ARIKAYCE.

Nephrotoxicity was observed during the clinical trials of ARIKAYCE in patients with MAC lung disease but not at a higher frequency than background regimen alone. Nephrotoxicity has been associated with the aminoglycosides. Close monitoring of patients with known or suspected renal dysfunction may be needed when prescribing ARIKAYCE.

Neuromuscular Blockade: Patients with neuromuscular disorders were not enrolled in ARIKAYCE clinical trials. Patients with known or suspected neuromuscular disorders, such as myasthenia gravis, should be closely monitored since aminoglycosides may aggravate muscle weakness by blocking the release of acetylcholine at neuromuscular junctions.

Embryo-Fetal Toxicity: Aminoglycosides can cause fetal harm when administered to a pregnant woman. Aminoglycosides, including ARIKAYCE, may be associated with total, irreversible, bilateral congenital deafness in pediatric patients exposed *in utero*. Patients who use ARIKAYCE during pregnancy, or become pregnant while taking ARIKAYCE should be apprised of the potential hazard to the fetus.

Contraindications: ARIKAYCE is contraindicated in patients with known hypersensitivity to any aminoglycoside.

Most Common Adverse Reactions: The most common adverse reactions in Trial 1 at an incidence ≥5% for patients using ARIKAYCE plus background regimen compared to patients treated with background regimen alone were dysphonia (47% vs 1%), cough (39% vs 17%), bronchospasm (29% vs 11%), hemoptysis (18% vs 13%), ototoxicity (17% vs 10%), upper airway irritation (17% vs 2%), musculoskeletal pain (17% vs 8%), fatigue and asthenia (16% vs 10%), exacerbation of underlying pulmonary disease (15% vs 10%), diarrhea (13% vs 5%), nausea (12% vs 4%), pneumonia (10% vs 8%), headache (10% vs 5%), pyrexia (7% vs 5%), vomiting (7% vs 4%), rash (6% vs 2%), decreased weight (6% vs 1%), change in sputum (5% vs 1%), and chest discomfort (5% vs 3%).

Drug Interactions: Avoid concomitant use of ARIKAYCE with medications associated with neurotoxicity, nephrotoxicity, and ototoxicity. Some diuretics can enhance aminoglycoside toxicity by altering aminoglycoside concentrations in serum and tissue. Avoid concomitant use of ARIKAYCE with ethacrynic acid, furosemide, urea, or intravenous mannitol.

Overdosage: Adverse reactions specifically associated with overdose of ARIKAYCE have not been identified. Acute toxicity should be treated with immediate withdrawal of ARIKAYCE, and baseline tests of renal function should be undertaken. Hemodialysis may be helpful in removing amikacin from the body. In all cases of suspected overdosage, physicians should contact the Regional Poison Control Center for information about effective treatment.

U.S. INDICATION

LIMITED POPULATION: ARIKAYCE® is indicated in adults, who have limited or no alternative treatment options, for the treatment of *Mycobacterium avium* complex (MAC) lung disease as part of a combination antibacterial drug regimen in patients who do not achieve negative sputum cultures after a minimum of 6 consecutive months of a multidrug background regimen therapy. As only limited clinical safety and effectiveness data for ARIKAYCE are currently available, reserve ARIKAYCE for use in adults who have limited or no alternative treatment options. This drug is indicated for use in a limited and specific population of patients.

This indication is approved under accelerated approval based on achieving sputum culture conversion (defined as 3 consecutive negative monthly sputum cultures) by Month 6. Clinical benefit has not yet been established. Continued approval for this indication may be contingent upon verification and description of clinical benefit in confirmatory trials.

Limitation of Use: ARIKAYCE has only been studied in patients with refractory MAC lung disease defined as patients who did not achieve negative sputum cultures after a minimum of 6 consecutive months of a multidrug background regimen therapy. The use of ARIKAYCE is not recommended for patients with non-refractory MAC lung disease.

Patients are encouraged to report negative side effects of prescription drugs to the FDA. Visit **www.fda.gov/medwatch**, or call 1-800-FDA-1088. You can also call the Company at 1-844-4-INSMED.

Please see Full Prescribing Information.

About Insmed

Insmed Incorporated is a global biopharmaceutical company on a mission to transform the lives of patients with serious and rare diseases. Insmed's first commercial product is a first-in-disease therapy approved in the United States, Europe, and Japan to treat a chronic, debilitating lung disease. The Company is progressing a robust pipeline of investigational therapies targeting areas of serious unmet need, including neutrophil-mediated inflammatory diseases and rare pulmonary disorders. Insmed is also advancing an early-stage research engine encompassing a wide range of technologies and modalities, including artificial intelligence-driven protein engineering, gene therapy, and protein manufacturing. Insmed is headquartered in Bridgewater, New Jersey, with additional offices and research locations throughoutthe United States, Europe, and Japan. Visit www.insmed.com to learn more.

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: failure to continue to successfully commercialize ARIKAYCE, our only approved product, in the US, Europe or Japan (amikacin liposome inhalation suspension, Liposomal 590 mg Nebuliser Dispersion, and amikacin sulfate inhalation drug product, respectively), or to maintain US, European or Japanese approval for ARIKAYCE; uncertainties or changes in the degree of market acceptance of ARIKAYCE by physicians, patients, third-party payors and others in the healthcare community; our inability to obtain full approval of ARIKAYCE from the FDA, including the risk that we will not successfully or in a timely manner validate a patient reported outcome (PRO) tool and complete the confirmatory post-marketing clinical trial required for full approval of ARIKAYCE; inability of us, PARI or our other third-party manufacturers to comply with regulatory requirements related to ARIKAYCE or Lamira® ; our inability to obtain and maintain adequate reimbursement from government or third-party payors for ARIKAYCE or acceptable prices for ARIKAYCE; development of unexpected safety or efficacy concerns related to ARIKAYCE, brensocatib, TPIP or our other product candidates; inaccuracies in our estimates of the size of the potential markets for ARIKAYCE, brensocatib, TPIP or our other product candidates or in data we have used to identify physicians, expected rates of patient uptake, duration of expected treatment, or expected patient adherence or discontinuation rates; the risks and uncertainties associated with, and the perceived benefits of, our secured senior loan with certain funds managed by Pharmakon and our royalty financing with OrbiMed, including our ability to maintain compliance with the covenants in the agreements for the senior secured loan and royalty financing and the impact of the restrictions on our operations under these agreements; our inability to create or maintain an effective direct sales and marketing infrastructure or to partner with third parties that offer such an infrastructure for distribution of ARIKAYCE or any of our product candidates that are approved in the future; failure to obtain regulatory approval to expand ARIKAYCE's indication to a broader patient population; risk that brensocatib or TPIP does not prove to be effective or safe for patients in ongoing and future clinical studies, including, for brensocatib, the ASPEN study; risk that our competitors may obtain orphan drug exclusivity for a product that is essentially the same as a product we are developing for a particular indication; failure to successfully predict the time and cost of development, regulatory approval and commercialization for novel gene therapy products; failure to successfully conduct future clinical trials for ARIKAYCE, brensocatib, TPIP and our other product candidates and our potential inability to enroll or retain sufficient patients to conduct and complete the trials or generate data necessary for regulatory approval of our product candidates or to permit the use of ARIKAYCE in the broader population of patients with MAC lung disease, among other things; risks that our clinical studies will be delayed, that serious side effects will be identified during drug development, or that any protocol amendments submitted will be rejected; risks that interim or partial data sets are not representative of a complete or larger data set or that blinded data will not be predictive of unblinded data; failure to obtain, or delays in obtaining, regulatory approvals for ARIKAYCE outside the US, Europe or Japan, or for our product candidates in the US,Europe, Japan or other markets, including separate regulatory approval for Lamira® in each market and for each usage; failure of third parties on which we are dependent to manufacture sufficient quantities of ARIKAYCE or our product candidates for commercial or clinical needs, to conduct our clinical trials, or to comply with our agreements or laws and regulations that impact our business or agreements with us; our inability to attract and retain key personnel or to effectively manage our growth; our inability to successfully integrate our recent acquisitions and appropriately manage the amount of management's time and attention devoted to integration activities; risks that our acquired technologies, products and product candidates are not commercially successful; inability to adapt to our highly competitive and changing environment; inability to access, upgrade or expand our technology systems or difficulties in updating our existing technology or developing or implementing new technology; risk that we are unable to maintain our significant customers; risk that government healthcare reform materially increases our costs and damages our financial condition; business or economic disruptions due to catastrophes or other events, including natural disasters or public health crises; risk that our current and potential future use of artificial intelligence and machine learning may

not be successful; deterioration in general economic conditions in the US, Europe, Japan and globally, including the effect of prolonged periods of inflation, affecting us, our suppliers, third-party service providers and potential partners; inability to adequately protect our intellectual property rights or prevent disclosure of our trade secrets and other proprietary information and costs associated with litigation or other proceedings related to such matters; restrictions or other obligations imposed on us by agreements related to ARIKAYCE or our product candidates, including our license agreements with PARI and AstraZeneca AB, and failure to comply with our obligations under such agreements; the cost and potential reputational damage resulting from litigation to which we are or may become a party, including product liability claims; risk that our operations are subject to a material disruption in the event of a cybersecurity attack or issue; our limited experience operating internationally; changes in laws and regulations applicable to our business, including any pricing reform and laws that impact our ability to utilize certain third parties in the research, development or manufacture of our product candidates, and failure to comply with such laws and regulations; our history of operating losses, and the possibility that we never achieve or maintain profitability; goodwill impairment charges affecting our results of operations and financial condition; inability to repay our existing indebtedness and uncertainties with respect to our ability to access future capital; and delays in the execution of plans to build out an additional third-party manufacturing facility approved by the appropriate regulatory authorities and unexpected expenses associated with those plans. 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, 2023 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.

With respect to the blended and blinded data observed from the ongoing TPIP study in PAH noted above, the dose titration and efficacy analyses were based on data available as of April 1, 2024, and the safety analyses were based on data available as of January 25, 2024, respectively. These findings may not be representative of results after the study is completed and all data are collected and analyzed. As a result, later interim data readouts and final data from this study may be materially different than the observations described above, including with respect to efficacy, safety and tolerability of TPIP.

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