ARIKAYCE®
(amikacin liposome inhalation suspension)
590 mg/8.4 mL

Limited Population

This handout provides distribution information for ARIKAYCE® (amikacin liposome inhalation suspension).

Insmed does not recommend the use of any particular specialty pharmacy

Not actual size.

The following specialty pharmacies are authorized to dispense ARIKAYCE

Contracted specialty pharmacy of record	Phone	Website
Kroger Specialty Pharmacy	855-274-1694	www.krogerspecialtypharmacy.com
Maxor Specialty (IV Solutions/ Pharmaceutical Specialties)	800-658-6046	www.maxor.com
Orsini Pharmaceutical Service, Inc.	800-672-0869	www.orsinihealthcare.com
PANTHERx Specialty Pharmacy, LLC	855-726-8479	www.pantherspecialty.com

Sample billing codes

The coding information discussed in this document is provided for informational purposes only, is subject to change, and should not be construed as legal advice. The codes listed herein may not apply to all patients or to all health plans. Conversely, additional codes not listed in this guide may apply to some patients. Providers should follow payer-specific coding requirements and exercise independent clinical judgment when selecting codes and submitting claims to accurately reflect the services and products furnished to a specific patient.

ICD-10-CM codes[1,2]

NTM lung disease	A31.0	Pulmonary mycobacterial infection
	A31.9	Mycobacterial infection, unspecified

CPT code[3]

Inhaler technique	94664	Demonstration and/or evaluation of inhaler techniques

CPT=*Current Procedural Terminology*; ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification*; NTM=nontuberculous mycobacterial.

INDICATION AND IMPORTANT SAFETY INFORMATION

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.

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.

Please see additional Important Safety Information on next page and full Prescribing Information, including Boxed Warning.

insmed®

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Trade unit product information[4]

Product name	ARIKAYCE
Generic name	amikacin liposome inhalation suspension
WAC	$10,164
NDC	71558-590-28
Trade unit dimensions	13.5"L x 8.5"W x 3.5"H
Trade unit weight	3.1 lb
Package size	28 vials
Dosage form	Inhalation suspension

NDC=National Drug Code; WAC=wholesale acquisition cost.

Each kit will have a unique US Drug Supply Chain Security Act–compliant 2-dimensional data matrix bar code with Global Trade Item Number, serial number, lot number, and expiration date. In addition, each case will have its own 2-dimensional data matrix bar code to identify the 6 kits inside.

Shipper case information

The initial patient shipment is 2 cartons. The first includes the Lamira™ Nebulizer System composed of an eBase® controller, an Aerosol Head, a Lamira Nebulizer Handset, a power cord, and accessories. This is delivered only once. The second (and all subsequent shipments) contains a 28-day supply of medication (28 vials), 1 Lamira Nebulizer Handset, and 4 Lamira Aerosol Heads.

Case quantity	6 trade units per case
Case weight	19.7 lb
Case dimensions	14.188"L x 17.688"W x 11.5"H

For more information, please call the *Arikares™ Support Program* at 1-833-ARIKARE (1-833-274-5273)

IMPORTANT SAFETY INFORMATION (cont'd)

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.

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.

References: 1. 2018 ICD-10-CM diagnosis code A31.0. ICD10Data.com website. https://www.icd10data.com/ICD10CM/Codes/A00-B99/A30-A49/A31-/A31.0. Accessed September 20, 2018. **2.** 2018 ICD-10-CM diagnosis code A31.9. ICD10Data.com website. https://www.icd10data.com/ICD10CM/Codes/A00-B99/A30-A49/A31-/A31.9. Accessed September 20, 2018. **3.** American Association for Respiratory Care. Coding guidelines for certain respiratory care services–January 2018. https://www.aarc.org/wp-content/uploads/2014/10/aarc-coding-guidelines.pdf. Accessed September 20, 2018. **4.** ARIKAYCE [package insert]. Bridgewater, NJ: Insmed Incorporated; 2018.

Please see full Prescribing Information, including Boxed Warning.

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