

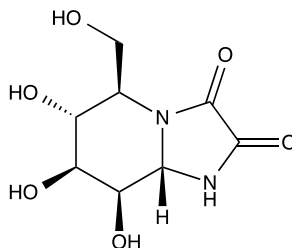
**Catalog # 10-2860**

**Kifunensine**

109944-15-2

(5*R*,6*R*,7*S*,8*R*,8*aS*)-Hexahydro-6,7,8-trihydroxy-5-(hydroxymethyl)-imidazo[1,2-*a*]pyridine-2,3-dione; FR-900494

Lot # X101395



Historically isolated from *Kitasatosporia kifunense*.<sup>1</sup> Inhibitor of class I  $\alpha$ -mannosidases which inhibit glycoprotein processing. Inhibits human endoplasmic reticulum  $\alpha$ -1,2-mannosidase I and Golgi Class I mannosidases IA, IB and IC with  $K_i$  values of 130 and 23 nM respectively.<sup>2</sup> Inhibition of endoplasmic reticulum  $\alpha$ -mannosidase I activity rescues the human  $\alpha$ -sarcoglycan R77C mutation suggesting a new pharmacological approach for limb girdle muscular dystrophy type 2D patients carrying mutations that impair  $\alpha$ -sarcoglycan trafficking.<sup>3</sup> Improves maturation of misfolded proteins.<sup>4</sup>

- 1) Iwami *et al.* (1987), *A new immunomodulator, FR-900494: taxonomy, fermentation, isolation, and physico-chemical and biological characteristics*; J.Antibiot. (Tokyo) **40** 612
- 2) Elbein *et al.* (1990), *Kifunensine, a potent inhibitor of the glycoprotein processing mannosidase I*; J.Biol.Chem. **265** 15599
- 3) Bartoli *et al.* (2008), *Mannosidase I inhibition rescues the human alpha-sarcoglycan R77C recurrent mutation*; Hum.Mol.Genet. **17** 1214
- 4) Wang *et al.* (2011), *Inhibition of endoplasmic reticulum-associated degradation rescues native folding in loss of function protein misfolding diseases*; J.Biol.Chem. **286** 43454

**PHYSICAL DATA**

Molecular Weight:	232.19
Molecular Formula:	C <sub>8</sub> H <sub>12</sub> N <sub>2</sub> O <sub>6</sub>
Purity:	>98% by TLC
	NMR: (Conforms)
Solubility:	DMSO (up to 35 mg/ml)
Physical Description:	White solid
Storage and Stability:	Store as supplied desiccated at -20°C for up to 1 year from the date of purchase. Solutions in DMSO may be stored at -20°C for up to 1 month.

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