COMPOUND GUIDE

SS-31 — Elamipretide / Forzinity (MTP-131)

Mitochondria-targeting peptide · FDA approved Sept 2025 (Barth syndrome) · SubQ injection

Quick Start

FDA APPROVED — NARROW INDICATION

SS-31 / Elamipretide was FDA approved on **September 19, 2025** under the brand name **Forzinity** for Barth syndrome in patients weighing ≥30 kg. This is a rare mitochondrial cardiomyopathy affecting mostly males. All other uses of SS-31 are investigational. Forzinity (280mg/3.5mL; 80mg/mL) is a ready-to-use solution — no reconstitution needed.

WHAT IT IS

SS-31 (also known as Elamipretide, MTP-131, or Bendavia) is a cell-permeable, mitochondria-targeting tetrapeptide. It concentrates at the inner mitochondrial membrane by binding cardiolipin — a phospholipid critical for cristae structure and electron transport chain function. It was developed by Stealth BioTherapeutics.

APPROVED DOSING (FORZINITY / BARTH SYNDROME)

40 mg SubQ once daily (patients ≥30 kg). Severe renal impairment (eGFR <30): reduce to 20 mg/day. Forzinity is supplied as a ready-to-use injection — no reconstitution needed. Community research protocols typically use 1–10 mg/day (far below the approved clinical dose).

REGULATORY STATUS

FDA approved (Forzinity) September 19, 2025 for Barth syndrome. All other indications are investigational. Not on WADA prohibited list. Research vials (lyophilised) require reconstitution and are for investigational use only.

Dosing Context

CONTEXT	DOSE	FREQUENCY	SOURCE
Forzinity (Barth syndrome)	40 mg (≥30 kg); 20 mg (eGFR <30)	Once daily SubQ	FDA-approved label
TAZPOWER trial	40 mg/day	Once daily SubQ	Crossover RCT; 12 weeks; primary endpoint missed
Community research	1–10 mg/day	Once daily SubQ	Off-label; not clinically validated

Supplies & Reconstitution (Research Vials)

Reconstitution Math — 10 mg Vial (3 mL BAC)

TARGET DOSE	UNITS (U-100)	DOSES / VIAL
2.5 mg	**75.0 units**	4
5 mg	**150.0 units**	2
10 mg	**300.0 units**	1

Reconstitute the 10 mg vial with **3.0 mL bacteriostatic water** → 3.33 mg/mL. U-100 syringe = 100 units per mL. Swirl gently — do not shake. Refrigerate 2–8°C.

Reconstitution Steps

01 **Warm the vial**
Allow vial to reach room temperature from freezer (10–15 min) before reconstitution.

02 **Swab both stoppers**
Alcohol swab both vial tops. Air-dry before inserting needle.

03 **Draw BAC water and inject along wall**
Draw required BAC water and run slowly down the inside glass wall of the peptide vial. Do not jet onto the powder.

Swirl gently until clear

04 Gently swirl — do not shake or vortex. Solution should be clear and colourless.

05 Refrigerate
Store at 2–8°C for up to 30 days. Protect from light. Do not freeze after reconstitution.

Mechanism of Action

CARDIOLIPIN BINDING

SS-31 selectively accumulates at the inner mitochondrial membrane by binding cardiolipin — a unique phospholipid found almost exclusively at IMM. Cardiolipin is critical for cristae curvature and electron transport chain (ETC) complex assembly.

CRISTAE STABILISATION

By binding cardiolipin, SS-31 stabilises IMM cristae structure. In Barth syndrome, mutant tafazzin causes abnormal cardiolipin remodelling and cristae fragmentation — SS-31 partially compensates for this structural deficit.

ETC SUPPORT / ATP PRODUCTION

Stabilised cristae support optimal assembly and function of ETC Complexes I–IV and ATP synthase (Complex V). Improved ETC efficiency supports ATP production in energy-depleted mitochondria — the primary clinical rationale in Barth syndrome.

ROS SCAVENGING

Reduces mitochondrial reactive oxygen species (ROS) production by improving ETC electron flow. Excess electron leak to oxygen produces superoxide — SS-31's ETC stabilisation reduces this leak, studied in ischaemia-reperfusion models.

Key Research

STUDY	DESIGN	KEY FINDING
TAZPOWER (Phase 3)	Crossover RCT; 12 weeks; Barth syndrome; 40 mg/day	Primary endpoint (6-min walk test) missed. 168-week OLE showed functional improvement.
HARP trial	Phase 2; heart failure; 40 mg/day 4 weeks	No significant improvement in 6-min walk vs placebo; well tolerated
Multiple ischaemia studies	Phase 2; renal ischaemia, AMI	Mostly negative or neutral primary endpoints; well-tolerated safety profile across trials
FDA approval (2025)	Barth syndrome; unmet need pathway	Approved September 19, 2025 as Forzinity based on OLE functional improvement data

Storage

FORM	TEMPERATURE	DURATION	NOTES
Forzinity (ready-to-use)	2–8°C (refrigerator)	Per label expiry	Do not freeze; protect from light; ready to inject — no reconstitution
Research vial (lyophilised)	−20°C (freezer)	Up to 24 months	Protect from light and moisture
Reconstituted research vial	2–8°C (refrigerator)	Up to 30 days	Do not freeze after reconstitution