

---

---

**UNITED STATES  
SECURITIES AND EXCHANGE COMMISSION  
WASHINGTON, D.C. 20549**

---

**SCHEDULE 13D**

**Under the Securities Exchange Act of 1934  
(Amendment No. 1)\***

---

**Milestone Pharmaceuticals Inc.**

(Name of Issuer)

**Common Shares, no par value per share  
(Title of Class of Securities)**

**59935V107  
(CUSIP Number)**

**Peter Haahr  
Novo Holdings A/S  
Tuborg Havnevej 19  
Hellerup, Denmark DK-2900  
+45 3527 6592**

*Copy to:*

**B. Shayne Kennedy, Esq.  
Latham & Watkins LLP  
650 Town Center Drive, 20<sup>th</sup> Floor  
Costa Mesa, CA 92626  
Telephone: (714) 540-1235**

(Name, Address and Telephone Number of Person Authorized to Receive Notices and Communications)

**September 18, 2019  
(Date of Event which Requires Filing of this Statement)**

---

If the filing person has previously filed a statement on Schedule 13G to report the acquisition which is subject of this Schedule 13D, and is filing this statement because of Rule 13d-1(e), 13d-1(f) or 13d-1(g), check the following box.

---

**Note:** Schedules filed in paper format shall include a signed original and five copies of the schedule, including all exhibits. See Rule 13d-7(b) for other parties to whom copies are to be sent.

---

\* The remainder of this cover page shall be filled out for a reporting person's initial filing on this form with respect to the subject class of securities, and for any subsequent amendment containing information which would alter disclosures provided in a prior cover page.

The information required on the remainder of this cover page shall not be deemed to be "filed" for the purpose of Section 18 of the Securities Exchange Act of 1934 (the "Act") or otherwise subject to the liabilities of that section of the Act but shall be subject to all other provisions of the Act (however, see the Notes).

---

---

1.	Name of Reporting Person: Novo Holdings A/S	
2.	Check the Appropriate Box if a Member of Group (See Instructions): (a) <input type="checkbox"/> (b) <input type="checkbox"/>	
3.	SEC Use Only:	
4.	Source of Funds: WC	
5.	Check if Disclosure of Legal Proceedings is Required Pursuant to <u>Items 2(d)</u> or <u>2(e)</u> : <input type="checkbox"/>	
6.	Citizenship or Place of Organization: Denmark	
Number of Shares Beneficially Owned By Each Reporting Person With:	7.	Sole Voting Power: 2,674,470
	8.	Shared Voting Power: 0
	9.	Sole Dispositive Power: 2,674,470
	10.	Shared Dispositive Power: 0
11.	Aggregate Amount Beneficially Owned by Each Reporting Person: 2,674,470	
12.	Check if the Aggregate Amount in Row (11) Excludes Certain Shares: <input type="checkbox"/>	
13.	Percent of Class Represented By Amount In Row (11): 10.92% (1)	
14.	Type of Reporting Person: CO	

- (1) Based upon 24,490,742 of the Issuer's Common Shares outstanding as of August 7, 2019 as reported in the Issuer's quarterly report (Form 10-Q) filed with the Securities and Exchange Commission ("SEC") on August 13, 2019.

This amendment (“Amendment No. 1”), amends the Schedule 13D originally filed with the SEC on May 14, 2019 (the “Schedule”) to remove reference to Nilesch Kumar, Ph.D. as serving as a member of the board of directors of the Issuer, update the directors and executive officers of the Reporting Person on Schedule I, and to report a change in the percentage of the Issuer’s Common Stock owned by the Reporting Person. All capitalized terms contained herein but not otherwise defined shall have the meanings ascribed to such terms in the Schedule.

**Item 2. Identity and Background**

- (a) Novo Holdings A/S is a Danish limited liability company that is wholly owned by Novo Nordisk Foundation (the “Foundation”), a Danish foundation. Novo Holdings A/S is the holding company in the group of Novo companies (currently comprised of Novo Nordisk A/S, Novozymes A/S and NNIT A/S) and is responsible for managing the Foundation’s assets, including its financial assets. Based on the governance structure of Novo Holdings A/S and the Foundation, the Foundation is not deemed to have any beneficial ownership of the securities of the Issuer held by Novo Holdings A/S. The members of the Novo Holdings Board of Directors are not deemed to beneficially own the securities held by Novo Holdings. Nilesch Kumar, Ph.D. is employed as a partner at Novo Ventures (US), Inc. and served on the Issuer’s board of directors from July 2017 until September 18, 2019. Dr. Kumar was not deemed to beneficially own the securities held by Novo Holdings.

The name of each director and executive officer of both Novo Holdings A/S and the Foundation is set forth on Schedule I to this Schedule 13D.

- (b) The business address of both Novo Holdings A/S and the Foundation is Tuborg Havnevej 19, 2900 Hellerup, Denmark.

The residence or business address of each director and executive officer of both Novo Holdings A/S and the Foundation is set forth on Schedule I to this Schedule 13D.

- (c) Novo Holdings A/S, a holding company that is responsible for managing the Foundation’s assets, provides seed and venture capital to development stage companies and invests in well-established companies within the life science and biotechnology sector.

The Foundation is a Danish self-governing and profit-making foundation, whose objectives are to provide a stable basis for commercial and research activities undertaken by the group of Novo companies and to support scientific, humanitarian and social purposes through grants.

- (d) Within the last five years, neither Novo Holdings A/S, the Foundation, nor any person named in Schedule I has been convicted in any criminal proceedings.

- (e) Within the last five years, neither Novo Holdings A/S, the Foundation, nor any person named in Schedule I was a party to a civil proceeding of a judicial or administrative body of competent jurisdiction and as a result of such proceeding was or is subject to a judgment, decree or final order enjoining future violations of, or prohibiting or mandating activities subject to, federal or state securities laws or finding any violation with respect to such laws.

**Item 5. Interest in Securities of the Issuer**

- (a) Novo Holdings A/S beneficially owns 2,674,470 Common Shares (the “Novo Shares”) representing approximately 10.92% of the Issuer’s outstanding Common Shares, based upon 24,490,742 of the Issuer’s Common Shares outstanding as of August 7, 2019 as reported in the Issuer’s quarterly report (Form 10-Q) filed with the SEC on August 13, 2019.

---

(b) Novo Holdings A/S is a Danish limited liability company wholly owned by the Novo Nordisk Foundation. Novo Holdings A/S, through its Board of Directors (the “Novo Board”), has the sole power to vote and dispose of the Novo Shares. The Novo Board may exercise voting and dispositive control over the Novo Shares with approval by a majority of the Novo Board. As such, no individual member of the Novo Board is deemed to hold any beneficial ownership or reportable pecuniary interest in the Novo Shares.

(c) Novo Holdings A/S has not effected any transactions in the Issuer’s ordinary shares within the past 60 days and neither the Foundation nor any person listed on Schedule I has effected any transactions in the Issuer’s ordinary shares within the past 60 days.

(d) Novo Holdings A/S does not know of any other person having the right to receive or the power to direct the receipt of dividends from, or the proceeds from the sale of the Issuer’s ordinary shares held in the name of the Novo Holdings A/S and reported herein.

(e) Not applicable.

---

**SIGNATURE**

After reasonable inquiry and to the best of my knowledge and belief, I certify that the information set forth in this statement is true, complete and correct.

Dated: September 20, 2019

Novo Holdings A/S

/s/ Peter Haahr

By: Peter Haahr

Its: Chief Financial Officer

---

**Schedule I**

Information regarding each director and executive officer of both Novo Holdings A/S and the Novo Nordisk Foundation is set forth below.

<i>Novo Holdings A/S</i>			
<b>Name, Title</b>	<b>Address</b>	<b>Principal Occupation</b>	<b>Citizenship</b>
Lars Rebién Sørensen, Chairman of the Board	Christianholms Tværvej 27 2930 Klampenborg Denmark	Professional Board Director	Denmark
Steen Riisgaard, Vice Chairman of the Board	Hestetangsvej 155, 3520 Farum, Denmark	Professional Board Director	Denmark
Jean-Luc Butel, Director	235 Arcadia Road, #03-04, 28984 Singapore	Global Healthcare Advisor, President, K8 Global Pte Ltd.	Singapore
Jeppe Christiansen, Director	Kollemosevej 37, 2830 Virum, Denmark	Chief Executive Officer, Fondsmaeglerselskabet Maj Invest A/S	Denmark
Francis Michael Cyprian Cuss, Director	111 Rippling Brook Way, Bernardsville, NJ 07924 USA	Former Executive Vice President and Chief Scientific Officer of Bristol-Myers Squibb	United Kingdom
Viviane Monges, Director	Chemin de Craivavers 32, 1012 Lausanne, Switzerland	Professional Board Director	France
Kasim Kutay, Chief Executive Officer of Holdings A/S	Bredgade 63, 3.th. 1260 Copenhagen K. Denmark	Chief Executive Officer of Novo Holdings A/S	United Kingdom
Peter Haahr, Chief Financial Officer of Novo Holdings A/S	Ordrup Have 21 2900 Charlottenlund Denmark	Chief Financial Officer of Novo Holdings A/S	Denmark

<i>Novo Nordisk Foundation</i>			
<b>Name, Title</b>	<b>Address</b>	<b>Principal Occupation</b>	<b>Citizenship</b>
Lars Rebién Sørensen, Chairman of the Board	Christianholms Tværvej 27 2930 Klampenborg Denmark	Professional Board Director	Denmark
Marianne Philip, Vice Chairman of the Board	Tranegårdsvej 5 2900 Hellerup Denmark	Attorney	Denmark
Steen Riisgaard, Director	Hestetangsvej 155 3520 Farum Denmark	Professional Board Director	Denmark
Birgitte Nauntofte, Chief Executive Officer	Engbakkevej 24 2920 Charlottenlund Denmark	Chief Executive Officer, Novo Nordisk Foundation	Denmark

---

*Novo Nordisk Foundation*

<b>Name, Title</b>	<b>Address</b>	<b>Principal Occupation</b>	<b>Citizenship</b>
Niels Peder Nielsen, Deputy CEO	Winthersvej 10, 3480 Fredensborg Denmark	Deputy CEO, Novo Nordisk Foundation	Denmark
Anne Marie Kverneland, Director	Nybrovej 216 2800 Kgs. Lyngby Denmark	Laboratory technician, Novo Nordisk A/S	Denmark
Lars Bo Køppler, Director	Anemonevej 7 3550 Slangerup Denmark	Technician, Novozymes A/S	Denmark
Lars Fugger, Director	72 Staunton Road, Headington OX3 7TP Great Britain	Professor, John Radcliffe Hospital, University of Oxford, Oxford, Great Britain	Denmark
Lars Henrik Munch, Director	Galionsvej 46 1437 København K Denmark	Professional Board Director	Denmark
Mads Boritz Grøn, Director	Horsevænget 4 3400 Hillerød Denmark	Senior Lead Auditor	Denmark
Liselotte Højgaard, Director	Grønningen 21 1270 København K Denmark	Professor	Denmark