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

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**FORM 6-K**

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**Report of Foreign Private Issuer  
Pursuant to Rule 13a-16 or 15d-16  
of the Securities Exchange Act of 1934**

September 25, 2017

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**PROQR THERAPEUTICS N.V.**

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2333 CK Leiden  
The Netherlands  
Tel: +31 88 166 7000**

(Address, Including ZIP Code, and Telephone Number, Including Area Code, of Registrant's Principal Executive Offices)

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Indicate by check mark whether the registrant files or will file annual reports under cover of Form 20-F or Form 40-F.

Form 20-F

Form 40-F

Indicate by check mark if the registrant is submitting the Form 6-K in paper as permitted by Regulation S-T Rule 101(b)(1):

Indicate by check mark if the registrant is submitting the Form 6-K in paper as permitted by Regulation S-T Rule 101(b)(7):

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On September 25, 2017, ProQR Therapeutics N.V. (“ProQR” or the “Company”) (NASDAQ: PRQR), issued a press release titled, “ProQR Announces Positive Top-Line Results from a Phase 1b Study of QR-010 in Subjects with Cystic Fibrosis.” A copy of this press release is furnished as Exhibit 99.1 to this Report on Form 6-K and is incorporated herein by reference. The Company hereby incorporates by reference the information contained herein into the Company’s registration statement on Form F-3 (File No. 333-207245).

**SIGNATURES**

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned, thereunto duly authorized.

**PROQR THERAPEUTICS N.V.**

Date: September 25, 2017

By: /s/ Smital Shah  
Smital Shah  
Chief Financial Officer

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**INDEX TO EXHIBITS**

**Number**

**Description**

99.1 Press Release of ProQR Therapeutics N.V., dated September 25, 2017, titled “ProQR Announces Positive Top-Line Results from a Phase 1b Study of QR-010 in Subjects with Cystic Fibrosis.”



FINAL – FOR RELEASE

# ProQR Announces Positive Top-Line Results from a Phase 1b Study of QR-010 in Subjects with Cystic Fibrosis

## Key updates

- QR-010 was observed to be safe and well-tolerated across all doses in this trial with no serious adverse events related to treatment.
- A clinically meaningful improvement of CF respiratory symptoms, as measured by CFQ-R RSS, was observed in 3 out of 4 multiple dose groups with a mean improvement of 13.0 to 19.2 points compared to placebo. In a pre-defined subgroup of subjects with a lower lung function at baseline, the mean improvement was up to 27.5 points compared to placebo.
- Magnitude of the benefit observed in CFQ-R RSS for these dose groups exceeded the established minimal clinically important difference of 4.0 points.
- In some multiple dose groups a supportive trend of improved lung function was observed up to 4.0% mean absolute change in ppFEV<sub>1</sub> compared to placebo. In a pre-defined subgroup of subjects with a lower lung function at baseline a mean absolute change in ppFEV<sub>1</sub> was observed up to 10.9% compared to placebo.
- After inhaled administration in some dose groups, QR-010 was detected in the blood.
- ProQR and the Cystic Fibrosis Foundation Therapeutics intend to expand their partnership to explore the inhaled oligonucleotide platform to target stop-codon mutations in CF.
- Management will discuss the top-line results during a conference call today at 5 pm ET.

LEIDEN, the Netherlands, September 25, 2017—ProQR Therapeutics N.V. (Nasdaq:PRQR) today announced positive preliminary top-line results from a Phase 1b safety and tolerability clinical trial (Study PQ-010-001; [NCT02532764](#)) of QR-010, a novel investigational RNA therapeutic in subjects with cystic fibrosis (CF). Full data from the trial will be presented at the North American CF Conference (NACFC) on November 2-4, 2017.

Study PQ-010-001 was a Phase 1b, randomized, double-blind, placebo-controlled, dose escalation study to evaluate the safety, tolerability and pharmacokinetics of QR-010 in adult subjects with CF homozygous for the F508del mutation. This trial studied 4 dose levels of QR-010 administered via inhalation in 4 single-dose and 4 multiple-dose groups. A number of exploratory efficacy endpoints were assessed in the multiple dose groups: respiratory symptoms (as measured by a validated patient-reported outcome tool, the Cystic Fibrosis Questionnaire-Revised Respiratory Symptom Score, or CFQ-R RSS), lung function (as measured by mean absolute change in percent predicted forced expiratory volume in 1 second, or ppFEV<sub>1</sub>), sweat chloride test and weight change. This study included subjects with, on average, a normal lung function at baseline (mean ppFEV<sub>1</sub> 86%, range 69-116%). As therapeutic trials typically study subjects with normal-to-severe lung function at baseline (ppFEV<sub>1</sub> <90%), a subgroup was pre-defined to analyze the exploratory efficacy endpoints in this population. The trial recruited 70 participants and was conducted at 23 sites in 10 countries in Europe and North America.

J. Stuart Elborn, the principal investigator of the study, Clinical Chair in Respiratory Medicine at Imperial College, Consultant at Royal Brompton Hospital, and immediate past-president of the European Cystic Fibrosis Society added, “QR-010 exceeded expectations in this study as an innovative investigational therapy for the treatment of cystic fibrosis for which the need remains high. The improvements demonstrated in reduction of

respiratory symptoms are very encouraging and intriguing and of course of enormous importance to people with CF. The results of this study together with the previous proof of concept study are strongly supportive of the further development of QR-010.”

### Top-Line Results

In this trial QR-010 was observed to be safe and well-tolerated across all doses, with an overall safety profile similar to placebo. There were no serious adverse events related to treatment. After inhaled administration in some dose groups, QR-010 was detected in the blood. Subjects who received QR-010 in the 6.25, 12.5 and 25 mg multiple dose groups reported fewer respiratory symptoms after 4 weeks of treatment as measured by the increased CFQ-R RSS, with mean improvements of 13.0, 19.2 and 14.3 points, respectively, compared to placebo. The effect was more pronounced in the pre-defined subgroup of subjects with a lower lung function at the start of the study (baseline ppFEV<sub>1</sub> 70-90%) with a mean increase of up to 27.5 points compared to placebo. These improvements exceeded the minimal clinically important difference (MCID) of 4.0 points. A supportive trend of improved lung function was observed in the same multiple dose groups, as measured by mean absolute change in ppFEV<sub>1</sub> compared to placebo. The trend was stronger in the subgroup of subjects with a lower lung function at baseline. The table below summarizes the data per multiple dose group. As expected, no effect was observed on sweat chloride and weight.

Groups (12 doses over 4 weeks)	n	Per protocol population				Pre-defined subgroup of subjects with baseline ppFEV <sub>1</sub> 70-90%				
		CFQ-R RSS		ppFEV <sub>1</sub>		CFQ-R RSS		ppFEV <sub>1</sub>		
		mean change vs placebo (p-value)	95% CI	mean absolute % change vs placebo (p-value)	95% CI	mean change vs placebo (p-value)	95% CI	mean absolute % change vs placebo (p-value)	95% CI	
6.25 mg	6	+13.0 (0.0585)	-0.5 ; 26.4	+1.2 (0.7266)	-6.0 ; 8.4	3	+23.2 (0.0315)	2.4 ; 44.1	+8.0 (0.1613)	-3.6 ; 19.5
12.5 mg	6	+19.2 (0.0072)	5.7 ; 32.7	+4.0 (0.2626)	-3.2 ; 11.2	4	+27.5 (0.0095)	7.9 ; 47.1	+10.9 (0.0461)	0.2 ; 21.6
25 mg	6	+14.3 (0.0399)	0.7 ; 27.9	-0.2 (0.9664)	-7.3 ; 7.1	5	+20.3 (0.0334)	1.9 ; 38.7	+4.7 (0.3410)	-5.5 ; 14.8
50 mg	5	+3.5 (0.6182)	-10.7 ; 17.6	-0.6 (0.8749)	-8.2 ; 7.0	4	+10.9 (0.2463)	-8.4 ; 30.2	+3.7 (0.4745)	-7.0 ; 14.3
Placebo	8	-6.5	-15.3 ; 2.4	-0.8	-5.5 ; 3.9	4	-11.8	-25.5 ; 2.0	-3.8	-11.3 ; 3.8

The Phase 1b study achieved its goals for evaluation of QR-010 including demonstrating safety and tolerability across a range of doses, identified a dosing window, exhibited uptake of the RNA oligonucleotide into circulation following inhalation, and demonstrated early signals of clinical efficacy.

“The positive results in the first two clinical trials of QR-010 significantly increase the confidence that QR-010 has the potential to become an effective treatment of CF. The improvement in CFQ-R RSS with the supportive FEV<sub>1</sub> data as shown in this Phase 1b trial is very exciting,” said Noreen R. Henig, M.D, Chief Medical Officer at ProQR. “I want to thank the entire CF community including those living with CF, clinical investigators, scientists, the CF Foundation, the European CF Society and the US and European therapeutic development networks for their unwavering commitment.”

### Partnership with Cystic Fibrosis Foundation Therapeutics (CFFT)

ProQR and CFFT entered into a partnership in 2014 to develop QR-010 for people with CF due to the F508del mutation. The initial partnership included support for the Phase 1b trial as well as the NPD proof of concept study that reported positive results in 2016. Based on the results of the clinical trials of QR-010, ProQR and CFFT intend to expand the partnership to explore the inhaled oligonucleotide platform for stop-codon mutations (also called “Class I” mutations) in CFTR. Stop-codon mutations cannot be targeted with small molecule potentiator or corrector molecules, and therefore have a high unmet medical need. ProQR intends to target these mutations using its proprietary Axiomer® technology, which has shown compelling data in non-clinical studies, to repair the stop-codon mutations in the RNA, leading to removal of the premature stop-

codon. Approximately 12,000 patients, accounting for 15% of CF patients in the western world, have a stop-codon mutation leading to a severe form of CF.

”The results of this Phase 1b trial support QR-010’s potential to be a meaningful therapy for people with cystic fibrosis,” said Daniel A. de Boer, Chief Executive Officer at ProQR. ”With these results in hand we are designing a path forward for the development of QR-010 either independently or with a potential partner. Furthermore we are looking forward to expanding our partnership with the CFFT to explore the inhaled oligonucleotide platform also for people that have CF due to stop-codon mutations.”

### **About the PQ-010-001 Trial**

The Phase 1b study was a trial designed to assess safety, tolerability and pharmacokinetics of QR-010. A number of exploratory efficacy endpoints were assessed in the multiple dose groups. A total of 4 dose levels were studied: 6.25, 12.5, 25 and 50 mg of QR-010 in solution per dose administered via inhalation using the PARI eFlow® nebulizer. Subjects eligible to participate were males and females of 18 years and over with a ppFEV<sub>1</sub> of  $\geq 70\%$  at time of inclusion, homozygous for the F508del mutation, and not taking CFTR modulator drugs. The study design planned to enroll 8 cohorts of 8 subjects (6 receiving QR-010, 2 receiving placebo). In cohorts 1-4, a single dose of QR-010 was administered, and in cohorts 5-8 twelve doses of QR-010 were administered over a 4-week period.

### **QR-010 Milestones**

- Technology for QR-010 in-licensed from Massachusetts General Hospital in 2012.
- Partnership with the CFF established to develop QR-010 for patients with the F508del mutation.
- *In vitro* proof of concept in three F508del CF assays.
- *In vivo* proof of concept in two assays, including nasal potential difference (NPD).
- Pre-clinical *in vitro* and *in vivo* proof of concept established for efficient inhaled delivery to the CF diseased lung in collaboration with the University of North Carolina at Chapel Hill.
- QR-010 granted fast-track status by the FDA and orphan drug designation from FDA and the European Commission.
- Program received funding from the European Union’s Horizon 2020 research and innovation programme.
- Clinical trial PQ-010-002 top-line data shows significant improvement of CFTR function as measured by NPD in subjects homozygous for the F508del mutation following topical administration of QR-010.
- Grant of two key patents, protecting QR-010 in the US and Europe.
- Preliminary top-line data from clinical trial PQ-010-001 shows QR-010 is detected in the blood after inhaled administration, was observed to be safe and well-tolerated and shows signals of efficacy.
- Full data for PQ-010-001 will be presented at the NACFC (November 2-4, 2017).

## Conference Call and Webcast Information

ProQR will host a conference call and webcast today at 5:00 PM Eastern Time (ET) or 11:00 PM Central European Time (CET). The conference call will be webcast live and a link to the webcast can be accessed through ProQR's website ([www.proqr.com](http://www.proqr.com)) and in the "Investors" section under "Events and Presentations". To ensure a timely connection, it is recommended that users register at least 15 minutes prior to the scheduled webcast. An archived webcast will be available on the company's website. To participate in the conference call, please dial in 5-10 minutes prior to start time and reference **Conference ID 8468989**:

Country	Toll Free	Direct
US	1 877 280 2296	+1 646 254 3365
Netherlands	0800 020 2576	+31 (0) 20 713 2789
United Kingdom	0800 279 4977	+44 (0) 20 3427 1919
Germany	0800 589 2674	+49 (0) 69 2222 10620
Belgium	0800 58032	+32 (0) 2 400 3463
Switzerland	0800 345 603	+41 (0) 44 580 7214
France	0805 631 580	+33 (0) 1 7677 2222

### About QR-010

QR-010 is a first-in-class RNA-based oligonucleotide designed to address the underlying cause of the disease by targeting the mRNA in CF patients that have the F508del mutation. The technology was exclusively licensed from Massachusetts General Hospital. The F508del mutation results in the production of a misfolded CFTR protein that does not function normally. QR-010 is a single agent designed to bind to the defective CFTR mRNA and to restore CFTR function. QR-010 is designed to be self-administered via an optimized eFlow® Nebulizer (PARI Pharma GmbH). eFlow® is a small, handheld aerosol delivery device which nebulizes QR-010 into a mist inhaled directly into the lungs. QR-010 has been granted orphan drug designation in the United States and the European Union and fast-track status by the FDA. The QR-010 project received funding from the European Union's Horizon 2020 research and innovation programme under grant agreement No 633545.

### About Cystic Fibrosis

Cystic fibrosis (CF) is the most common fatal inherited disease in the Western world and affects over 75,000 patients worldwide. In people with CF, a defective CFTR gene causes a thick buildup of mucus in the lungs, pancreas and other organs. In the lungs, the mucus clogs the airways and traps bacteria leading to infections, extensive lung damage and eventually, respiratory failure. There is no cure for CF. Disease manifestations lead to a shortened life expectancy with a median age of death of 30 years. Although over 1,900 CF-causing gene mutations have been identified, approximately 85% of all CF patients are affected by the F508del mutation. Among all CF patients, approximately 50% are homozygous for the F508del mutation.

### About ProQR

ProQR Therapeutics is dedicated to changing lives through the creation of transformative RNA medicines for the treatment of severe genetic rare diseases such as cystic fibrosis, Leber's congenital amaurosis 10 and dystrophic epidermolysis bullosa. Based on our unique proprietary RNA repair platform technologies we are growing our pipeline with patients and loved ones in mind.

\*Since 2012\*



## FORWARD-LOOKING STATEMENTS

This press release contains forward-looking statements. All statements other than statements of historical fact are forward-looking statements, which are often indicated by terms such as “anticipate,” “believe,” “could,” “estimate,” “expect,” “goal,” “intend,” “look forward to”, “may,” “plan,” “potential,” “predict,” “project,” “should,” “will,” “would” and similar expressions. Forward-looking statements are based on management’s beliefs and assumptions and on information available to management only as of the date of this press release. These forward-looking statements include, but are not limited to, statements regarding QR-010, including its clinical development and therapeutic potential, including future development plans, and statements regarding the partnership with CFFT. Our actual results could differ materially from those anticipated in these forward-looking statements for many reasons, including, without limitation, risks associated with our clinical development activities, including that positive results observed in our prior and ongoing studies may not be replicated in later trials or guarantee approval of any product candidate by regulatory authorities, that a Fast Track designation by the FDA may not actually lead to a faster development, regulatory review or approval process, and that we may not be able to realize the potential benefits of orphan drug exclusivity, manufacturing processes and facilities, regulatory oversight, product commercialization, intellectual property claims, and the risks, uncertainties and other factors in our filings made with the Securities and Exchange Commission, including certain sections of our annual report filed on Form 20-F. Given these risks, uncertainties and other factors, you should not place undue reliance on these forward-looking statements, and we assume no obligation to update these forward-looking statements, even if new information becomes available in the future.

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