



(51) International Patent Classification:

A61K 31/00 (2006.01) **A61K 31/519** (2006.01)
A61K 31/416 (2006.01) **A61K 31/53** (2006.01)
A61K 31/437 (2006.01) **A61K 31/5377** (2006.01)
A61K 31/4418 (2006.01) **A61P 21/00** (2006.01)
A61K 31/4439 (2006.01) **G01N 33/15** (2006.01)
A61K 31/496 (2006.01)

(21) International Application Number:

PCT/US2018/054642

(22) International Filing Date:

05 October 2018 (05.10.2018)

(25) Filing Language:

English

(26) Publication Language:

English

(30) Priority Data:

62/568,673	05 October 2017 (05.10.2017)	US
62/568,754	05 October 2017 (05.10.2017)	US
62/682,565	08 June 2018 (08.06.2018)	US
62/682,563	08 June 2018 (08.06.2018)	US

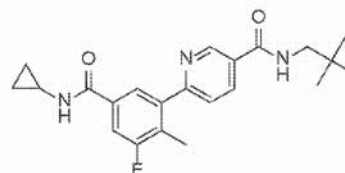
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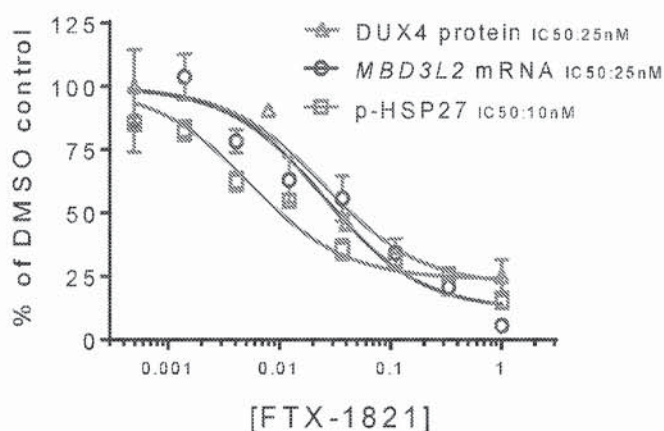
(81) Designated States (unless otherwise indicated, for every kind of national protection available): AE, AG, AL, AM, AO, AT, AU, AZ, BA, BB, BG, BH, BN, BR, BW, BY, BZ, CA, CH, CL, CN, CO, CR, CU, CZ, DE, DJ, DK, DM, DO, DZ, EC, EE, EG, ES, FI, GB, GD, GE, GH, GM, GT, HN, HR, HU, ID, IL, IN, IR, IS, JO, JP, KE, KG, KH, KN, KP, KR, KW, KZ, LA, LC, LK, LR, LS, LU, LY, MA, MD, ME,

(54) Title: P38 KINASE INHIBITORS REDUCE DUX4 AND DOWNSTREAM GENE EXPRESSION FOR THE TREATMENT OF FSHD



FTX-1821

FIG. 7



(57) Abstract: The disclosure relates to methods and compositions including p38 kinase inhibitors and agents that regulate expression of DUX4 and downstream genes including but not restricted to ZSCAN4, LEUTX, PRAMEF2, TRIM43, MBD3L2, KHDCIL, RFPL2, CCNA1, SLC34A2, TPRX1, PRAMEF20, TRIM49, PRAMEF4, PRAME6, PRAMEF15, or ZNF280A. Methods useful for treating a disease associated with abnormal DUX4 and downstream gene expression (e.g., Fascioscapulohumeral muscular dystrophy) are disclosed.

MG, MK, MN, MW, MX, MY, MZ, NA, NG, NI, NO, NZ,
OM, PA, PE, PG, PH, PL, PT, QA, RO, RS, RU, RW, SA,
SC, SD, SE, SG, SK, SL, SM, ST, SV, SY, TH, TJ, TM, TN,
TR, TT, TZ, UA, UG, US, UZ, VC, VN, ZA, ZM, ZW.

- (84) Designated States** (*unless otherwise indicated, for every kind of regional protection available*): ARIPO (BW, GH, GM, KE, LR, LS, MW, MZ, NA, RW, SD, SL, ST, SZ, TZ, UG, ZM, ZW), Eurasian (AM, AZ, BY, KG, KZ, RU, TJ, TM), European (AL, AT, BE, BG, CH, CY, CZ, DE, DK, EE, ES, FI, FR, GB, GR, HR, HU, IE, IS, IT, LT, LU, LV, MC, MK, MT, NL, NO, PL, PT, RO, RS, SE, SI, SK, SM, TR), OAPI (BF, BJ, CF, CG, CI, CM, GA, GN, GQ, GW, KM, ML, MR, NE, SN, TD, TG).

Declarations under Rule 4.17:

- *as to applicant's entitlement to apply for and be granted a patent (Rule 4.17(ii))*

Published:

- *with international search report (Art. 21(3))*
- *before the expiration of the time limit for amending the claims and to be republished in the event of receipt of amendments (Rule 48.2(h))*
- *with sequence listing part of description (Rule 5.2(a))*

From the INTERNATIONAL BUREAU

PCTNOTIFICATION OF THE RECORDING
OF A CHANGE(PCT Rule 92bis.1 and
Administrative Instructions, Section 422)

To:

FARMER, Dean
COOLEY LLP
1299 Pennsylvania Ave. Suite 700
Washington, District of Columbia 20004
ÉTATS-UNIS D'AMÉRIQUE

Date of mailing (day/month/year) 22 July 2019 (22.07.2019)	IMPORTANT NOTIFICATION
Applicant's or agent's file reference FULC02602WO	
International application No. PCT/US2018/054642	International filing date (day/month/year) 05 October 2018 (05.10.2018)

1. The following indications appeared on record concerning: ☐ the applicant ☒ the inventor ☐ the agent ☐ the common representative		
Name and Address	State of Nationality	State of Residence
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	Facsimile No.	
	E-mail address	
2. The International Bureau hereby notifies the applicant that the following change has been recorded concerning: ☐ the person ☐ the name ☐ the address ☐ the nationality ☐ the residence		
Name and Address SHEN, Ning 82 Hancock Street, Apt. 1 Cambridge, MA 02139 United States of America	State of Nationality	State of Residence
	Telephone No.	
	Facsimile No.	
	E-mail address ☐ Notifications by e-mail authorized	
3. Further observations, if necessary: The person identified in Box 2 has been added as inventor only.		
4. A copy of this notification has been sent to: ☒ the receiving Office ☐ the International Preliminary Examining Authority ☒ the International Searching Authority ☒ the designated Offices concerned ☐ the Authority(ies) specified for supplementary search ☐ the elected Offices concerned ☐ other:		

The International Bureau of WIPO 34, chemin des Colombettes 1211 Geneva 20, Switzerland	Authorized officer N'Diaye Anta e-mail pct.team6@wipo.int Telephone No. +41 22 338 74 06
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	Facsimile No.	
	E-mail address	
2. The International Bureau hereby notifies the applicant that the following change has been recorded concerning:		
☐ the person	☐ the name	☐ the address
☐ the nationality		
☐ the residence		
Name and Address CHANG, Aaron Nakwon 41 Preston Road Lexington, MA 02420 United States of America	State of Nationality	State of Residence
	Telephone No.	
	Facsimile No.	
	E-mail address ☐ Notifications by e-mail authorized	
3. Further observations, if necessary: The person identified in Box 2 has been added as inventor only.		
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☒ the International Searching Authority	☒ the designated Offices concerned	
☐ the Authority(ies) specified for supplementary search	☐ the elected Offices concerned	
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	Facsimile No.	
	E-mail address	
2. The International Bureau hereby notifies the applicant that the following change has been recorded concerning:		
☐ the person	☐ the name	☐ the address
☐ the nationality		
☐ the residence		
Name and Address RONCO, Lucienne, V. 16 Garden Road Wellesley, MA 02481 United States of America	State of Nationality	State of Residence
	Telephone No.	
	Facsimile No.	
	E-mail address ☐ Notifications by e-mail authorized	
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Name and Address ROBERTSON, Alan Scott 160 Cambridgepark Drive, Apt. 278 Cambridge, MA 02139 United States of America	State of Nationality	State of Residence
	Telephone No.	
	Facsimile No.	
	E-mail address ☐ Notifications by e-mail authorized	
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