

MacMullen CM, Zhou Q, Snider KE, Tewson PH, Becker SA, Aziz AR, Ganguly A, Shyng S-L, Stanley CA. Diazoxide-unresponsive congenital hyperinsulinism in children with dominant mutations of the β -cell sulfonylurea receptor SUR1. *Diabetes* 2011;60:1797–1804

In the print version of the article listed above, one of the dominant diazoxide-responsive mutations reported by us (Pinney et al.) in 2008, R1539Q, was erroneously expressed as R1539E (this mutation has not yet been reported in an affected patient). These data were used in our article to compare surface expression and channel responses to MgADP and diazoxide with the dominant diazoxide-unresponsive mutations. Fig. 4, on page 1802, shows the data for R1539E that is labeled erroneously as R1539Q. After detecting the error, expression studies were carried out for the R1539Q mutation. Functionally, the R1539Q and R1539E mutations behave very similarly. We have added these data to a revised version of Fig. 4, shown below. The gray square labeled “H” shows the data for R1539E, while the hatched box labeled “K” shows the correct data for R1539Q. The value for surface expression (% of wild-type channel level) should be for R1539Q = 98.9 ± 12.58 versus R1539E = 72.6 ± 10.2 ; MgADP stimulation (% current) for the R1539Q = 5.54 ± 1.63 versus R1539E = 12.9 ± 4.3 ; and diazoxide stimulation (% current) for the R1539Q = 18.81 ± 6.11 versus R1539E = 35.1 ± 6.5 . The mean surface expression (% of wild-type channel level) for the group of dominant diazoxide-responsive SUR1 mutations is now 93.68 ± 5.07 ($n = 10$), with the data for R1539E replaced with R1539Q, versus 91.05 ± 5.67 ($n = 10$) as originally published in the article by Pinney et al. The means of MgADP and diazoxide stimulation (% current) (shown in Table 2 of our *Diabetes* article) for the dominant diazoxide-responsive SUR1 mutations (versus the values published in the article by Pinney et al.) should be 14.52 ± 5.15 ($n = 10$) (versus 15.26 ± 5.06 [$n = 10$]) and 18.33 ± 6.06 ($n = 10$) (versus 19.96 ± 6.29 [$n = 10$]), respectively. The P values for the means of MgADP and diazoxide stimulation when comparing the dominant diazoxide-unresponsive mutations to the dominant, diazoxide-responsive mutations are $P = 0.0003$ (versus $P = 0.0002$) and $P = 0.0002$ (versus $P = 0.0003$), respectively. Since the effect of the error is small, the conclusions stated in the article are not changed.

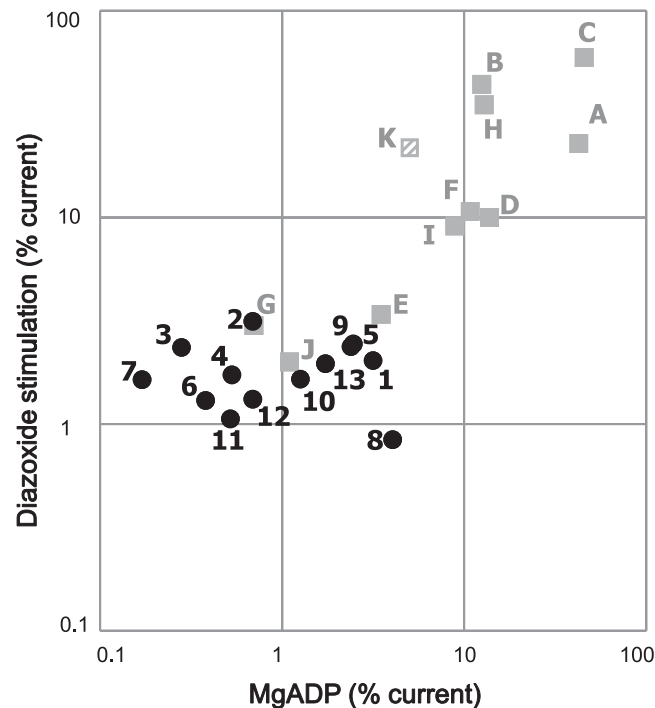


FIG. 4. Responsiveness of dominant diazoxide-unresponsive and dominant diazoxide-responsive SUR1 mutations. Responses to diazoxide and MgADP of expressed K_{ATP} channels containing diazoxide-unresponsive SUR1 mutations (black circles) and diazoxide-responsive SUR1 mutations (gray squares) are compared. Diazoxide-unresponsive mutations are labeled according to numbers in Table 2. Diazoxide-responsive mutations: A = D310N, B = R370G, C = R1353H, D = K1374R, E = G1478V, F = G1479R, G = S1386P, H = R1539E, I = I1512T, J = E1507K, and K = R1539Q.

The online version reflects these changes.