

Bolwetse jwa von Willebrand ke eng?



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Bolwetse ja Willebrand ke eng (VWD)?

von Willebrand disease (VWD) ke bolwetse ja go tswa madi mo go sa tlwaelesegang. Batho ba ba nang le VWD ba na le mathata a kotla e e agang mesifa e e thusang go laola go tshologa madi . Ga ba na selekanyo se se lekaneng sa kotla e e agang mesifa kgotsa e ba nang nayo ga e bereke jaaka e tshwanetse. Go tsaya lebaka gore madi a ipope matlhole le gore go tswa madi go eme.

Go na le mefuta e e farologaneng ya VWD. Mefuta yotlhe e bakiwa ke mathata a kotla ya von Willebrand factor (VWF). Fa tshika ya madi e golafala go bo go tswa madi, VWF e thusa dikarolo tsa madi tse gotweng di-platelets go kopana go dira lethole la madi le le emisang go tshologa madi.

VWD ke mofuta o o tlwaelesegileng thata wa go tswa madi mo go sa tlwaelesegang o batho ba nang nao. E ama banna le basadi. VWD ga e bogale thata jaaka makoa a mangwe a go tshologa madi. Batho ka bantsi ba ba nang le VWD ba ka nna ba bo ba sa itse gore ba na le yone ka gore dikai tsa bone tsa go tswa madi ga se tse di tshwenyang. Mo bathong ba bantsi ba ba nang le VWD, matshelo a bone ga a kgoreletsege ka tsela epe, kontle ga ba na le kgobalo e e tlhwaafetseng kgotsa ba tlhoka loaro. Le fa gontse jalo, mefuta yotlhe ya VWD e ka baka mathata a go tswa madi thata.

Batho ba tsenwa ke VWD jang?

Motho o tsholwa ka VWD. E fetela ko mothong ka segotso se ngwana a se tsayang go tswa mo motsading wa bong bongwe le bongwe. Fa gongwe go na le bosupi ja ditso tsa mathata a go tshologa madi mo lwapeng/lesikeng. Le fa gontse jalo, dikai tsa go tswa madi di kgona go farologana mo bathong mo lwapeng. Fa gongwe ga gona ditso tsa mathata a go tswa madi, bolwetse bo direga fela ka diphetogo tsa karolwana ya segotso ya VWD pele ngwana a tsholwa.

Dikai tsa VWD ke di fe?

Dikai tsa konokono tsa VWD ke:

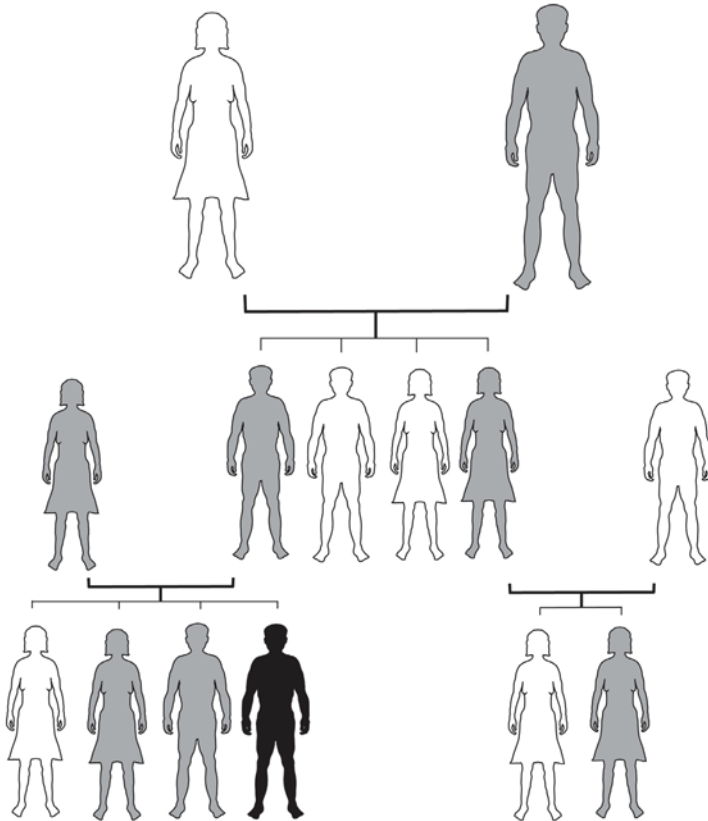
- Go nna le letsadi motlhofo/go rurubala
- Go tshabelelwa ke mokola(go tswa madi mo nkong) kgotsa mokola o tswa lebaka le le leele
- Go tswa madi mo marining
- Go tswa madi lebaka le leele mo dikgobalong tse di potlana
- setswalo se se bokete kgotsa se tsaya lebaka le le leele
- go tswa madi mo teng ga mpa
- Go tswa madi lebaka le le leele morago ga kgobalo, loaro, go ntshiwa leino kgotsa pelegi

Bontsi ja batho ba ba nang le VWD ga ba supe dikai dipe kgotsa ba itemogela dikai tse di seng kae. Batho ba ba nang le VWD e e tlhwaafetseng ba kgona go nna le mathata a go tswa madi a le mantsi. Dikae di kgona go fetoga mo tsamaong ya nako. Fa gongwe VWD e lemogiwa fa go setse go na le go tshologa madi thata morago ga kotsi e e masisi kgotsa tsamaiso ya loaro kgotsa go ntsha leino

Basadi ba bantsi ba supa dikai tsa VWD go na le banna. Basadi ba ba nang le VWD gantsi ba tswa madi thata kgotsa ba tsaya lebaka le le leele ba le mo setswalong le morago ga pelegi. Basadi bangwe ba ba nang le VWD ba itemogela ditlhabi tse di botlhoko thata ka nako ya setswalo kgotsa ga ba bone setswalo sa bone sentle.

Setlhopha sa madi se ka nna le seabe. Batho ba setlhopha sa madi sa O gantsi ba na selekanyo se se ko tlase sa VWD go na le batho ba setlhopha sa A, B kgotsa AB. Se se raya gore batho ba ba nang le VWD le setlhopha sa madi sa O ba ka nna le mathata a go tswa madi thata.

Go tsholwa ka von Willebrand disease



Inheritance of von Willebrand disease



NONE



MILD



SEVERE

VWD e lemogiwa jang?

Ga go motlhofo go lemoga kgotsa go tshwara VWD. Batho ba ba belaelang gore ba na le mathata a bolwetse ja go tswa madi ba tshwanetse go bona moitseanape wa mathata a madi. Ditlhatlhobo tse di maleba di ka dirwa ko lefelong la kalafi ya malwetse a go tswa madi. Ka gore kotla e e agang mesifa ya VWD e dira ditiro tse di fetang bongwe, go tshwanetse ga dirwa ditlhatlhobo tse di fetang bongwe go tlhola VWD

Ditlhatlhobo tsa laboratori tsa VWD di thata go dirwa. VWD ga e tsharwe ka ditlhatlhobo tsa madi tsa gale. Tlhatlhobo e akaretsa go meta selekanyo sa motho le go dira ga VWF, le sa mofuta mongwe wa kotla e e agang mesifa e e thusang madi gore a ipope matlhole ya factor VIII(FVIII). Tlhatlhobo gantsi e a boelediwa ka gore selekanyo sa motho sa VWF le FVIII di kgona go farologana ka di nako tse di farologaneng.

Mefuta e e farologaneng ya VWD

Go na le mefuta e le meraro ya VWD. Mo mofuteng o le mongwe wa VWD, bolwetse bo kgona go nna jo bo lotsatsa , jwa bokete jo bo fa gare kgotsa jo bo bogale. Dikai tsa go tswa madi di kgona go farologana go tswa fela gore VWF e dira jang. Go botlhokwa go itse mofuta wa VWD o motho a nang nao, ka gore kalafi e farologana ka mofuta mongwe le mongwe.

Mofuta wa ntlha(Type 1) wa VWD: ke one o o bonalang kgotsa o o tlwaelesegileng thata. Batho ba ba nang le Type 1 VWD ba na le selekanyo se se ko tlase sa VWF. Dikai gatsi ga se tse di reng sepe. Go a kgonega gore motho yo o nang le Type 1 VWD a itemegele go tswa madi mo go bokete.

Mofuta wa bobedi(Type 2) wa VWD: Go nna le bogole mo popegong ya VWF. Kotla ya e e agang mesifa ya VWF ga e bereke sentle e be e baka go wela tlase ga go dira ga VWF. Go na le mefuta e e farologaneng ya bogole ja VWD. Dikai gantsi ke tse di fa gare.

Mofuta wa boraro(Type 3) wa VWD: ke mofuta o o bogale wa VWD. Batho ba ba nang le Type 3 VWD ga ba na VWF gotlhelele kgotsa ke ya selekanyo se se nnyennyane. Dikai ke tse di maswe thata. Batho ba ba nang le Type 3 ba kgona go tswela madi mo mesifeng le ditokololo, fa gongwe ba sa gobatswa ke sepe

VWD e alafiwa jang?

VWD e kgona go alafiwa ka molemo wa desmopressin, ke motswako o o komota o o nang le karolwana e e thusang madi go ipopa matlhole o o nang le VWF, kgotsa ka melemo e mengwe e e laolang go tswa madi. Kalafi e e fiwang e laolwa ke mofuta wa VWD o motho a nang nao. Batho ba ba nang le mefuta e e bonolo ya VWD ga se gantsi ba tlhoka kalafi kontle ga ba ka nna le loaro kgotsa go ntsha leino.

Desmopressin e bereka sentle thata go alafa Type 1 VWD, mme e ka dirisiwa go go kganela go tswa madi mo mothong yo o nang le mongwe wa mefuta wa Type 2 VWD. E dirisiwa go laola go tswa madi ka nako ya thuso ya potlako kgotsa loaro. E kgona go tsewa e le mokento kgotsa e sunyediwa mo nkong ka marothodinyana gore e tsholetse selekanyo sa VWF le FVIII gore e thuse madi go ipopa matlhole. Desmopressin ga e bereke mo go mongwe le mongwe. Ngaka o tshwanetse go dira ditlathlho bo go bona gore a motho o kgona go tsibogela molemo. Ka tshwanelo, ditlathlho bo di tshwanetse go dirwa pele ga kalafi e tlhokega.

Metswako ya digatsetsamadi Metswako e e komota e e nang le karolwana e e thusang madi go ipopa matlhole (**Factor concentrates**) e dirisiwa fa molemo wa desmopressin o sa bereke sentle kgotsa fa diphatsa tsa go tshologa madi thata di le kwa godimo. Metswako eno e na le wa VWF le FVIII. Ke one mofuta wa kalafi o o siametseng Type 3 VWD, mefuta mengwe ya Type 2 VWD le go tshologa madi mo go gaketseng kgotsa loaro le le tona mo mefuteng yotlhe ya VWD.

Go dutla madi go tswa mo dikarolong di tshwana mo teng ga nko, legano, mala le popelo go ka laolwa ke melemo ya **tranexamic acid** (Cyklokapron), **aminocaproic acid** (Amicar), kgotsa **fibrin glue**. Le fa gontse jalo, tsone di dirisiwa go tshwara lethole gore le seka la thubega e seng go thusa gore go dira lethole.

Kalafi ya di-hormone jaaka tse di nowang tsa boiphemelo, di thusa go oketsa selekanyo sa VWF le FVIII le go laola madi ka

nako ya setswalo. Fa go sa dirisiwe kalafi ya di-hormone, metswako e e kganelang go thubega/kgaogana ga lethole la madi(Antifibrinolytic agents) e ka kgona go alafa setswalo se se bokete. Dikalafi tsa mofuta o di ka nna le ditlamorago, ka jalo, batho ba ba nang le VWD ba tshwanetse go bua le ngaka wa bone mabapi le ditlamorago tse di ka nnang teng.

Dikgang tse di amang basetsana le bomme ba ba nang le VWD

Bomme ba ba nang le VWD ba tshabelelwa ke go nna le dikai tsa bolwetse go na le borre ka mabaka a go tsena mo setswalong le pelegi. Basetsana ba kgona go nna le setswalo se se bokete fa ba simolola go thatswa. Bomme ba ba nang le VWD gantsi ba itemogela setswalo se se bokete ebile se tsaya lebaka. Go nna le setswalo se se bokete go ka baka go wela tlase ga madi mo mmeleng (bolwetse ja tlhaelo ya madi, go wela tlase ga kotla e e agang madi mo go bakang letsapa le go tlhoka nonofo mo mmeleng). Bomme ba ba nang le VWD ba tshwanetse go nna ba tlholwa kgapetsakgapetsa gore ga ba na bolwetse jwa tlhaelo ya madi.

Mme yo o nang le VWD o tshwanetse go bona ngaka ya bomme fela fa a lemoga gore o imile. Moitseanape wa pelegi o tshwanetse go dirisana le ba lefelo la kalafi ya mathata a go tswa madi gore a kgone go fa mme tlhokomelo e e maleba ka nako ya boimana le pelegi. Ka nako ya boimana, bomme ba itemogela koketsego ya selekanyo ya VWF le karolwana e e thusang madi go ipopa matlhole. Se se thusa go sireletsa go tshologa madi ka nako ya pelegi. Le fa gontse jalo, morago ga pelegi, selekanyo sa karolwana e e thusang madi go ipopa matlhole se a fokotsega mme bomme ba ba nang le VWD ba ka kgona go tshologa madi.

Bomme ba ba gaufi le go khutla ga go bona modikologo wa kgwedi, ba dingwaga tse 45 le 50 ba mo diphatseng tsa kgonagalo ya go tshologa madi. Go botlhokwa gore mme yo o nang le VWD a tsewelele a tshwaragane le ngaka ya gagwe ya malwetse a bomme fa a ntse a atamela go khutla ga go bona modikologo wa kgwedi .

Dintlha tse di botlhokwa go batho ba ba nang leVWD

- Nna o tshwere otshedimose tso ka ga bokoa jwa gago, mofuta wa kalafi e o e filweng ke ba botsogo, leina le mogala wa ngaka ya gago le lefelo le o alafiwang kwa go lone. . Ka nako tsa tshoganetso, leseka la bongaka kgotsa sesupo sengwe se ka supegetsa badiri ba botsogo gore o na le bothata jwa go tswa madi
- Neela dikolo tshedimose tso ka VWD le gore diemo di ka laolwa jang fa di ka tlhaga.. Go tswa mokola ke jone bothata jo bo tlwaelesegileng kwa dikoleng.
- Ikwadise ko lefelong le le itebagantseng le go tlhatlhoba le go alafa malwetse a go tswa madi, ka gore ba ka kgona go fa tlhokomelo ya maemo a ntlha
- Botsa ngaka ka melemo yotlhe. Tila melemo mengwe e e rekisiwang mo mabenkeleng ka gore e ka kgoreletsa go gatsela ga madi.
- Itshidile ka nako tso tle go nonotsha ditokololo le mesifa gore di nne ba itekanetse.
- Fa o tsaya mosepele, ba batle di aterese le megala ya mafelo a dithuso tsa malwetse a go tswa madi gore ba tsamaye ba di tshotse ko ba yang teng.

Selected resources

World Federation of Hemophilia

www.wfh.org

- The Basic Science, Diagnosis, and Clinical Management of von Willebrand Disease
- Pregnancy in Women with Inherited Bleeding Disorders
- Gynecological Complications in Women with Bleeding Disorders
- Desmopressin (DDAVP) in the Treatment of Bleeding Disorders: The First 20 Years

Canadian Hemophilia Society

www.hemophilia.ca

- All About von Willebrand Disease
- Amicar and Cyklokapron, A Guide for Patients and Caregivers
- Desmopressin, A Guide for Patients and Caregivers

U.S. National Hemophilia Foundation

www.hemophilia.org/resources/handi_pubs.htm

- von Willebrand Disease: Just the FAQs
- A Guide for Women and Girls with Bleeding Disorders
- Project Red Flag – www.projectredflag.org

Association française des hémophiles

www.afh.asso.fr

- La maladie de Willebrand
- www.orpha.net/data/patho/Pub/fr/Willebrand-FRfrPub3497.pdf

Schweizerische Hämophilie-Gesellschaft

www.shg.ch

- Formes particulières d'hémophilie, la maladie de von Willebrand
- Informationen zur Hämophilie, Von Willebrand Krankheit

Selected resources

Haemophilia Foundation Australia

www.haemophilia.org.au

- A Guide for People Living with von Willebrand Disorder
- Meeting von Willebrand Disorder for the First Time: A Guide for Parents
- Understanding von Willebrand Disorder: A Guide for Teachers

Angelo Bianchi Bonomi Haemophilia Thrombosis Centre

- von Willebrand Disease: A Complex, Not Complicated Disorder (If Known)

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