

Lip-kaak- en gehemeltespleten in Nederland

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Graag wil ik met dit artikel mijn kijk op schisis in Nederland weergeven, gebaseerd op literatuur en mijn eigen ervaring als plastisch chirurg in het Erasmus MC Sophia Kinderziekenhuis.

SCHISIS

Schisis is de Griekse term voor spleet en is bij velen bekend geworden door de lip-kaak-gehemeltespleet (cheilognathopalatoschisis). Om onderscheid te maken met andere vormen van schisis, zoals de gastroschisis, wordt het ook wel orofaciale schisis genoemd. Deze afwijking komt in Nederland voor bij ongeveer 1 op de 600 geboortes oftewel jaarlijks zo'n 350 kinderen bij 200.000 geboortes per jaar. Dit getal geldt voor het Kaukasische ras, bij het Indische ras komt het vaker voor, bij het Negroïde ras minder. Opvallend is dat de variatie in incidentie geldt voor de lipspleet al of niet gecombineerd met een kaak- en/of gehemeltespleet. De geïsoleerde gehemeltespleet heeft daarentegen dezelfde incidentie bij alle rassen (ongeveer 1 op 2500). De afwijking kan alleen aan de lip voorkomen, aan de lip en kaak, alleen aan het gehemelte of aan de lip/kaak en het gehemelte. De afwijking kan in het betreffende deel volledig aanwezig zijn (complete spleet) of er bestaat nog een verbinding tussen de spleetdelen (incomplete spleet). Als er slechts een dunne welkodelenverbinding is, dan spreken we van een 'simonartsbandje', is de verbinding vrijwel compleet met onregelmatigheid in het verbindingsvlak dan spreken we van een 'embryonaal litteken' oftewel een 'forme fruste'. Bij het gehemelte kan de spleet compleet zijn (harde en zachte deel) of incompleet. Er is een grote variatie in incomplete gehemeltespleten: 1) incompleet in het harde deel en compleet in het zachte deel, 2) niet in het harde deel maar compleet in het zachte deel, 3) incompleet in het zachte deel in alle lagen, 4) een submucoseuze gehemeltespleet dat wil zeggen de palatummusculatuur sluit in het midden niet aan terwijl het slijmvlies gesloten is met een gespleten huijgje (bifide uvula). 5) alleen een gespleten huijgje (bifide uvula).

DE KENMERKEN

Bij de complete enkelzijdige lip-kaak-gehemeltespleet zien we geen verbinding tussen de lipdelen ter hoogte van de philtrumwal (figuur 1a en 1b). De musculus orbicularis oris is op dat niveau ook onderbroken en volgt aan de laterale zijde de rand van de lip tot de basis van de neusleugel, aan de mediale zijde is de spier onderontwikkeld in het philtrum tot aan de spleet en eindigt in de basis van de columella,



Figuur 1a en 1b. Enkelzijdige lip-kaak-gehemeltespleet.

De spleet heeft de vorm van een omgekeerde V, waarbij de neusleugel de verbinding maakt tussen de beide zijden van de spleet. Aan de laterale zijde is de hoogte van de lip verminderd, aan de mediale zijde is het philtrum verkort, maar is de philtrumwal aan de gezonde zijde behouden evenals twee derde van Cupido's boog en het philtrumkuiltje. De premaxilla heeft geen verbinding met de laterale bovenkaak. Mediaal is de premaxilla naar buiten gedraaid en heeft projectie, lateraal is de maxilla onderontwikkeld en naar achteren verplaatst. De neus is door de wijkende delen van de spleet verbreed aan de aangedane zijde. Het neusseptum is caudaal uit de mediane botgroeve gedissocieerd richting de niet aangedane zijde. Hierbij is de columella aan de basis ook naar de niet aangedane zijde verplaatst. Het septum is convex naar de aangedane zijde daarbij de adenoweg belemmerend. Het 'lower lateral' kraakbeentje is aan de aangedane zijde onder spanning gekomen en wordt plat getrokken, waardoor de neustip aan de aangedane zijde verlaagd is met separatie van de 'dome'. Dit geeft een kortere columella aan

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