



Faculty of Family Planning and Reproductive Health Care Clinical Effectiveness Unit

A unit funded by the FFPRHC and supported by the University of Aberdeen to provide guidance on evidence-based practice

MEMBERS' ENQUIRY RESPONSE

Enquiry Reference: 1925

**Sent: 01 May 2007
Prepared: 01 May 2007**

A: Question

For a woman with Klippel-Trenaunay Weber Syndrome what types of contraception are suitable?

B: Response

Klippel-Trenaunay syndrome (KTS) is a congenital circulatory disorder characterised by hemangiomas (abnormal benign growths on the skin consisting of masses of blood vessels), arteriovenous abscesses, and varicose veins, usually on the limbs. The cause of the disorder is largely unknown. Most patients will demonstrate all 3 signs of the clinical syndrome: port-wine stain, varicose veins, and bony and soft tissue hypertrophies. Medical treatment is conservative and symptomatic and include pressure garments, for chronic venous insufficiency, lymphedema, recurrent cellulitis and recurrent bleeding from capillary or venous malformations of the extremity. Pain management is also often required. Cellulitis and thrombophlebitis can be managed with analgesics, elevation, antibiotics and corticosteroids. In patients with a history of cellulitis, intermittent or prophylactic antibiotics may be considered. Anticoagulant therapy is indicated in acute thrombosis and prophylactically prior to surgical procedures. Laser treatment of the hemangioma can be effective in lightening the colour of the port-wine stain.

The United Kingdom *Medical Eligibility Criteria for Contraceptive Use* (UKMEC) does not give any specific recommendations for women with Klippel-Trenaunay Weber Syndrome. However, it does give recommendations for venous thromboembolism (VTE) which are described in detail in the response below.

In summary, for a woman with KTS and a history of VTE she may choose progestogen-only pills, implants or injectables, a copper-bearing intrauterine device or a levonorgestrel-releasing intrauterine system. If she has current VTE (with the use of anticoagulants) then she may choose progestogen-only pills or barrier methods. Concomitant drug use (antibiotics) may contraindicate use in some circumstances.

C: Evidence-Based Medicine Question *(which guided our literature search strategy)*

Population: Women with Klippel-Trenaunay Weber syndrome

Intervention: Contraception

Outcome: Safety

Keywords: Klippel-Trenaunay Weber syndrome; contraception; safety; 1925

D: Information Sources

The CEU searched the following sources in developing this Member's Enquiry Response

Source Searched	Information Identified
Existing FFPRHC and RCOG guidance	See below
The National Guidelines Clearing House	No relevant information
The WHO <i>Improving Access To Quality Care In Family Planning. Medical Eligibility Criteria For Contraceptive Use 2004</i> and <i>Selected Practice Recommendations For Contraceptive Use, 2004</i>	No relevant information
The Cochrane Library	No relevant information
MEDLINE and EMBASE from 1996 to 2007	No relevant information

E: Evidence Reviewed

Background

Klippel-Trenaunay syndrome (KTS) is a congenital circulatory disorder characterised by hemangiomas (abnormal benign growths on the skin consisting of masses of blood vessels), arteriovenous abscesses, and varicose veins, usually on the limbs. The affected limbs may be enlarged and warmer than usual. Fused toes or fingers, or extra toes or fingers may be present. Bleeding may occur, often as a result of rectal or vaginal tumour. The cause of the disorder is largely unknown. Most patients will demonstrate all 3 signs of the clinical syndrome: port-wine stain, varicose veins, and bony and soft tissue hypertrophies.¹

Medical treatment is conservative and symptomatic and include pressure garments, for chronic venous insufficiency, lymphedema, recurrent cellulitis and current bleeding from capillary or venous malformations of the extremity. Pain management is also often required. Cellulitis and thrombophlebitis can be managed with analgesics, elevation, antibiotics and corticosteroids. In patients with a history of cellulitis, intermittent or prophylactic antibiotics may be considered. Anticoagulant therapy is indicated in acute thrombosis and prophylactically prior to surgical procedures. Laser treatment of the hemangioma can be effective in lightening the colour of the port-wine stain.²

FFPRHC publications

The United Kingdom *Medical Eligibility Criteria for Contraceptive Use* (UKMEC)³ does not give any specific recommendations for women with KTS. However, concerning venous thromboembolism (VTE) the UKMEC states that the use of combined hormonal contraception (pills, patch and ring) for those with a history of VTE or current VTE (on anticoagulants) is unacceptable health risk (UKMEC category 4).

Women with a history of VTE can use progestogen-only contraception (pills, implant or injectable) however, if they currently have VTE they may use progestogen-only pills (UKMEC 2 where the advantages of using the method outweigh the theoretical or proven risks) but use of progestogen-only implants or progestogen-only injectables should be used in conjunction with clinical judgement and may need appropriate specialist referral.

Women with a history of VTE can choose a copper-bearing intrauterine device (Cu-IUD) or a levonorgestrel-releasing intrauterine system (LNG-IUS) without restriction (UKMEC 1 and 2 respectively). Women with current VTE may consider the use of intrauterine devices (both Cu-IUD or LNG-IUS) but perhaps should consider delaying insertion until anti-coagulants have stopped due to potential risk of bleeding during the insertion procedure.

The table below highlights the UKMEC categories for contraceptive use for women with a history of or current VTE³

Venous thromboembolism (VTE)	CHC	POP	DMPA-NET-EN	IMP	Cu-IUD	LNG-IUS	Barrier methods
History of VTE	4	2	2	2	1	2	1
Current VTE (on anticoagulants)	4	2	3	3	3	3	1

Abbreviations: CHC – combined hormonal contraception; POP = progestogen-only pills; DMPA = depot medroxyprogesterone acetate; NET-EN = norethisterone enanthate; IMP = progestogen-only implants; Cu-IUD = copper-bearing intrauterine device; LNG-IUS = levonorgestrel-releasing intrauterine system.

Some antibiotics can have liver-enzyme inducing properties and therefore concomitant drug use of combined hormonal contraception; progestogen-only pills and progestogen-only implants should be avoided. Women with KTS should consider this when choosing a contraceptive.

For a woman with KTS and a history of VTE she may choose progestogen-only pills, implants or injectables, a copper-bearing intrauterine device or a levonorgestrel-releasing intrauterine system. If she has current VTE (with the use of anticoagulants) then she may choose progestogen-only pills or barrier methods. Concomitant drug use (antibiotics) may contraindicate use in some circumstances.

F: References

1. National Institute of Neurological Disorders. NINDS Klippel-Trenaunay Syndrome (KTS) information page. http://www.ninds.nih.gov/disorders/klippel_trenaunay/klippel_trenaunay.htm?css=print. 2007.
2. Lisko JH. Klippel-Trenaunay-Weber Syndrome. <http://www.emedicine.com/derm/topic213.htm>. 2007.
3. Faculty of Family Planning and Reproductive Health Care Clinical Effectiveness Unit. UK Medical Eligibility Criteria for Contraceptive Use. <http://www.ffprhc.org.uk/admin/uploads/UKMEC200506.pdf>. 2006.

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Enquiry response by LA