

Parental Knowledge of Congenital Adrenal Hyperplasia (CAH) Vietnam & Indonesia



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Introduction

 CAH is associated with significant physical and psychosocial consequences for affected children and families where access to treatment is suboptimal



In Summary

• These results highlight the importance of providing

- The Incidence of Congenital Adrenal Hyperplasia (CAH) in some Asian countries is far higher than in Australia (e.g. 1:6000 Filipino Newborn Screening Program)
- Necessary resources for families in LMICs in Asia are limited:-
 - Reliable access to affordable essential medicines is problematic
- Medical expertise is usually centralised in major tertiary referral hospitals
- Families often lack confidence in the care provided by local practitioners
- Educational resources on CAH in local languages (hard copy and online) can be limited
- Many families living remotely travel long distances for medical care and to purchase medication
- Low income families struggle to afford health care

Children in low- and middle-income countries (LMICs) are at especial risk of preventable morbidity and mortality associated with CAH.

Background

CLAN (Caring and Living as Neighbours - an Australian based NGO established in 2004) has been working with families and healthcare teams in Vietnam and Indonesia, to optimise quality of life for children living with chronic health conditions such as CAH.

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Hyperplasia (CAH) Knowledge	2. In CAH the body makes: a) too much cortisol and too much androgen b) too little cortisol and not enough androgen c) too little cortisol and too much androgen d) unsure	8. Fludrocortisone (Flor/nef) helps regulate: a) salt balance b) biodo sugar levels c) growth d) unsure	
Assessment Questionnaire	3. CAH is caused by: a) a virus b) a faufly gene c) a harmful environmental agent d) unsure 4. The chance that CAH will recur in a family is: a) one in foru with every pregnancy b) with every pregnancy	9. The main reason for treating CAH is to: a) promote normal growth and development b) prevent an adrenal crisis c) both a) and b) d) unsure 10. Treatment for CAH is required: a) until you grow out of it b) until adulthood c) lifelon	
	 c) never c) never d) unsure 5. To have CAH the child must inherit the gene from: a) both the mother and father b) the mother only c) the father only d) unsure 6. Cortisol is: a) an essential vitamin b) essential for idecreasing blood sugar c) a homone essential for life 	(a) unsure (a) unsure (b) unsure (c) unsure (c) unsure (c) unsure (c) an Adrenal Crisis someone urgently needs: (c) a hydrocortisone injection and fluids via a drip (c) a hydrocortisone injection and fluids via a drip (c) unsure (c) u	
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Với mỗi câu hỏi, chỉ khoanh tròn một câu trả (For each question, circle one answer only)	lời duy nhất.	Untuk setiap pertanyaan, harap lingkari satu jawaban (For each question, circle one answer only)	saja
1. Tăng sản Thượng thận Bắm sinh là một tính trạng của : a. tuyển yên b. tuyển thương thận c. bưông trừng hay tinh hoàn d. không biết	 Quá nhiều androgen có thể: làm chân tăng trường gây ra tăng trường nhanh chóng và dây thi sớm làm châm phát triển dây thì không biết 	 Tidak makan Hidrokortison menyebabkan: kadar gula darah rendah & tekanan darah rendah kurang energi dan mengantuk a) dan b) tidak yain 	 Pasien HAK dengan muntaber yang berkelanjutan perlu: mengulang dools Hidrokortison suntikan Hidrokortison, lalu pengi ke 85 minum calran manis-asin tidak yakin
 Trong Tăng sản Thượng thận Bẩm sinh cơ thế tạo ra: quá nhiều cortisol và quá nhiều androgen quá it cortisol và không đủ androgen quá it cortisol và quá nhiều androgen không biết 	 e. invorcevnesenie (reinfiner) gulg bleu noa a. c. din bång muldi b. mit c. då duröng huyét c. t. tång truðing d. khöng biết 	 rengewaturu windi Jefini Krok Kenlangan garam bisangk termasuk: Hidrokortison (Pyrone) dan Fludrokortison (Pyrone) saja Fludrokortison (Pyrone) saja Fludrokortison (Pyrone) saja Hidrakortison (Pyrone) saja 	 Pasien HAK mempunyai resike krisis adrenal saat mereka: berolahraga berolahraga menjalani anestesi (untuk operasi) menjalani ujian sekolah di tidak yakin 20. Suntikan Hidrokortishon harus diberikan: ke vena di lengan
 Tăng sản Thượng thận Bắm sinh bị gây ra bởi: một virút một virút virút virút một virút virút	 Lý do chính để điều trị Tăng sản Thượng thận Bảm sinh là: a. thức đảy tăng trưởng và phát triển bình thường ngăn ngừa con khủng hoàng thượng thần c. cả a và b không biết 	 a. segera makan obat a. segera makan obat b. melipat gandakan dosis selanjutnya c. tidak peri mekakan aga-apa & makan obat sepert biasa pada dosis selanjutnya d. tidak yakin 15. Kalau pashen HAK sakit dengan demam tinggi, anda haris: 	 b. teo tot lengan atau paha c. te bawah lapisan kulit lengan atau paha d. tidak yakin 21. Koonutukai medik untuk anak dan remaja dianjurkan untuk: a. şerahun tehali b. 2. kali stenham
 Khả năng Tăng sản Thượng thận Bắm sinh sẽ xuất hiện lại trong một gia đinh là: a. một phản trở cho một lần mang thai b. với mỗt lần mang thai c. không bag giớ d. không bắt 	 Điều trị Tăng sản Thượng thận Bắm sinh thì cản: a. cho đến khi ban hết tàng trưởng b. cho đến tuổi trưởng thành c. suốt đới 	a. medmästikain meressa borstistansis sampai meresia merasa labib baik b. menghubangi döhter beri Härdstorison dengan dösi lebih, lalu menghubangi dökter tidak yakin 16. Kalau pasien IAK sedikit pilek tapi sehat, anda	 report outris years examined tidak yakin Alasan utama mengenakan gelang atau kalun medis adalah karena ka: memberinah nama & alamat memberinah nama & alamat memberikan diagnosa & pengobatan daruat menupikkan kerabat terdekat anda
 Để mặc Tăng sản Thượng thận Bẩm sinh đứn bé cản thừa hưởng gen từ: a. cả bỏ và mẹ b. ch từ mẹ c. chi từ nẹ c. chi từ bỏ d. không biết 	 không biết 11. Trong cơn Khủng hoàng Thượng Thận bệnh nhân cản khân trương: a. uống Paracetamol (Panadol) b. uống muối, đường và nghi ngợi 	haras: a. meningkatkan dosis Hidrokortison b. mengurangkan dosis hidrokortison c. Memonitor: sulvu tubuh dan menguranati ketat d. tidak yakia 137. Kalau pasten HAK puscit dan mengantuk, mereka perlui:	d, tidak yakin
 Cortisol là: a. một vitamin thiết yếu b. cần thiết cho hạ lượng đường trong mâu c. một hoặc-mộn cần thiết cho sự sống thiết cho bắt 	 c. chích hydrocortisone và truyền dịch không biết 	a. dosis obat makan Hidrokortison lebih b. Suntikan Hidrokortison, lalu pergi ke RS c. Cairan manis-asin untuk diminum d. tidak yakin	

Results

Demographics of the 3 Centres:

Total No. Analysed	245	Female	Male	Ave Age Years	Average time to diagnosis	Classical CAH	Non Classical CAH/ Unsure Unknown	Hydrocortisone & Florinef/ Hydrocortisone only
Hanoi	155	79	76	8 (1-19 ^{yr})	<1 yr (1 @ 4yr)	102	19/12 Unknown = 16	H&FI = 93 H = 50 Unknown = 7
нсмс	66	33	33	5(19 ^{mth} -15 ^{yr})	<1 yr	34	6/14 Unknown = 12	$\begin{array}{ll} H\&FI &= 60\\ H &= 6\\ Unknown &= 3 \end{array}$
Jakarta	24	14	10	5 (4 ^{mth} -15 ^{yr})	< 1 yr	16	2/4 Unknown = 2	$\begin{array}{rrrr} H\& FI & = & 7\\ H & = & 7\\ Unknown & = & 0 \end{array}$

families of children living with CAH in LMICs with further education in a structured way. Children living with CAH have a right to fulfill their life potential, and we all have a collective responsibility to help them achieve that.

- The diagnosis of CAH can be emotionally devastating for any family, and can greatly influence the attainment of knowledge. Having some understanding about what may influence knowledge about CAH and its management is critical to improving outcomes for these families. These groups had a basic understanding of the basic pathophysiology of CAH except for some uncertainty about inheritance in Q4 & 5. There fact that clinical management seemed to challenge families could be influenced by local issues in relation to adequate supply of affordable medication. Families may be aware of the value of Florinef or the need to triple dose with hydrocortisone during times of stress, but avoid using it because of limited access to affordable supplies. A multi-sectorial approach to improving health outcomes is essential, and this is facilitated by CLAN's strategic "Five Pillar" framework for action.
- Paediatric centres in Vietnam and Indonesia see relatively high numbers of children with CAH compared to Australia, yet the numbers of specialist Paediatric Endocrinologists and specialist nurses are far less available. Many families live remotely and travel long distances to obtain medical care and medications, which can be expensive.
- The data set obtained for the Indonesian group who attended the CAH Club meeting in Jakarta (Kahaki) was indeed small. Indonesia is a country with a large island archipelago, making travel difficult and unaffordable for LMIC families. Attendance at such meetings can also be influenced by a significant need for privacy and confidentiality about health diagnoses. There can also be an element of denial and a reluctance to share experiences which can be culturally significant.

CLAN works in collaboration with a wide range of partners to support CAH Communities in LMICs so that the children might fulfill their right to health. CAH CLAN, and these have been made freely available to all families in Vietnam and Indonesia.





CAHKAQ knowledge scores:

CAH Knowledge	Mean	SD	Minimum score	Maximum score over total	% Less than mean	
Hanoi	30	+/-7.05	7/44	44/44	41.9% (n=65)	
НСМС	31	+/-7.0	9/44	44/44	45% (n=30)	
Jakarta	30	+/-8.54	20/44	39/44	37% (n= 9)	

Summary of knowledge deficits:

Knowledge difficulties	Percentage incorrect or unsure > 3 rd tertile	n=155 Hanoi %	n=66 HCMC %	n=24 Jakarta %	n=154 Australia %
Q4	The chance CAH will recur in a family is	44.20	39.39	41.66	17.3
Q9	The main reason for treating CAH	30.20	40.90	20.83	17.7
Q11	In an adrenal crisis someone urgently needs	16.10	30.30	2.94	3.8
Q12	Not taking Hydrocortisone causes	36.24	54.54	54.16	31.7
Q14	If a person misses a medication they should	41.61	45.45	70.83	30.8
Q16	If a person has slight cold they should	48.99	34.84	66.66	19.8
Q17	If a person is pale & extremely drowsy	54.36	51.51	79.16	31.4
Q18	A person with persistent diarrhea & vomiting	45.63	37.88	29.16	13.0
Q19	A person with CAH is at risk of an adrenal crisis	61.74	65.15	37.5	24.9
Q20	Hydrocortisone injection should be given into	68.45	71.21	75.0	24.3
Q21	Medical follow-up is recommended	61.74	91.21	37.5	38.9
Q22	Main reason for wearing a medical alert	44.96	51.51	37.5	4.9

The knowledge base of CAH families across the three Centres appears similar.

- CAH Support Groups facilitate education and psychosocial support for large numbers of families in a time-efficient way, although limited resources and funding for these activities are a constraining factor.
- Free online access to translated educational resources and psychosocial support for CAH families and health professionals will be an important innovation as Internet access increases in LMICs. Quality, validated educational material in a multimedia format will allow all families and health professionals to view, hear and revise key information as needed. Updating material regularly would be relatively cost-effective.
- The CAH Psychosocial Education Program (CAHPeP) has been developed particularly for this purpose. It has been translated into Vietnamese and Bahasa Indonesian (see Poster P03.20) and is accessible on the Internet at www.CAHPePTalk.com. Promoting use of these education materials amongst families and evaluating subsequent knowledge growth and satisfaction respectively will be an important next step.

Evaluate the knowledge of families attending the CAH Clan Club meetings in Vietnam and Indonesia using the CAHKAQ translated into both languages.

Method

- CAHKAQ was initially developed and validated in Australia¹. Translation was undertaken by health professionals experienced in CAH from both Vietnam and Indonesian hospitals.
- CAHKAQ includes demographics and treatment, with 11 questions on pathophysiology and 11 questions on CAH management.
- 3. CAHKAQ was given to each family at registration for a CAH Club Meeting. Additional clinical data was collected height, weight and blood pressure.

Parents in Vietnam and Indonesia appeared to have a reasonable knowledge about the basic pathophysiology of the condition (Q1-11), with some uncertainty about the inheritance of CAH (Q 4 & 5).

There was a significant deficit in the area of clinical management, and uncertainty about medication dosing. More than 50% of families did not understand the consequences of not taking or missing Hydrocortisone doses. Sick day management issues were a particular challenge for families in Vietnam and Indonesia (Q12-19), a finding similar to Australian data ². Like families in Australia, families living in Vietnam and Indonesia also struggled to identify features of serious illness and when to appropriately administer stress doses of hydrocortisone.

Questions (Q17, 18, 19, 20) relating to more severe illness had the highest incorrect answers (37.5 - 79.16%) and this was consistent across the three Centres.

Conclusion

Health education is an integral component in any health setting, however it should not occur in isolation. Using the CAHKAQ to evaluate knowledge provides important information on which to focus education strategies, as well as providing baseline data point for evaluating knowledge attainment and understanding. Education and understanding are important for enhancing health outcomes for all patients.

References:

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- 2. Irene Mitchelhill (2010) Evaluation of a psychosocial education program for Congenital Adrenal Hyperplasia. *Master of Nursing (Hons) Thesis*. University of Technology, Sydney.