different locations for its replication. The TuMV NIa protease is also responsible for the cleavage of the TuMV polyprotein at seven different locations. To determine the substrate specificity of the NS3 and NIa proteases, amino acid sequences cleaved by the NS3 and NIa proteases were obtained from randomized sequence libraries by using a screening method referred to as GASP. Based on statistical analyses of the obtained cleavable sequences, consensus substrate sequences were deduced: Gln-Glu-Thr-Leu-Val▽Ser for HGV NS3 protease and Yaa-Val-Arg-His-Gln▽Ser for TuMV Nia protease, with Yaa being one of aliphatic amino acids. The relevance of this peptide as a cleavable substrate was further supported by molecular modeling of the HGV NS3 protease.

F104 Molecular Characterization of Blue Pigment Binding Protein-1 from Pieris rapae.

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We cloned and molecularly characterized a blue pigment binding protein-1 (BP-1) cDNA from cabbage white butterfly. The cDNA has a length of 474 bp coding for a 158-residue protein with a predicted molecular mass of 18,051 Da. The calculated isoelectric point is 8.2. Multiple alignment analysis of amino acid sequence revealed that BP-1 is most similar to bilin-binding protein (BBP) from *Pieris brassicae* (93%) followed by BBP of *Galleria mellonella* (48.7%). The BP-1 transcript was detected by Northern blot analysis in fat body and midgut.

F105 Mutations of Methyl-CpG binding protein 2 gene in Rett syndrome patients

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Rett syndrome (RTT [OMIM#312750]) is an X-linked dominant neurodevelopmental disorder affecting 1/10,000 - 15,000 females. RTT results from mutations in *MeCP2*, methyl-CpG binding protein 2, in 80% of patients. *MeCP2* gene spans 76kb in Xq28 and encodes two functional domains, a methyl-binding domain (MBD) and transcriptional repression domain (TRD). Authors have already reported the results of mutation screening of *MeCP2* in 20 Korean patients with RTT. In this presentation, we will report the newly identified 10 mutations including five missense mutations (D97Y, L100V, R133C, T158M, R306C), four nonsense mutations (R168X, R255X, R270X, R294X), and one frameshift mutation (a 41-bp deletion at 1157~1197). Two of these (D97Y and L100V) were novel mutations. We also established 21 lymphoblastoid cell lines from RTT patients. These cell lines will be used as valuable materials to ascertain whether *MeCP2* is related to the X chromosome inactivation, if any, what is its function.

F106 Comparative analysis of deleted region at 7q11.23 in WS and SVAS patients

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Williams syndrome (WS) is a complex developmental disorder with multisystem manifestations including supravalvular aortic

stenosis (SVAS). Patients with WS show allelic loss of *ELN*, exhibiting a submicroscopic deletion at 7q11.23. While the involvement of elastin mutation in isolated SVAS is indisputable, the nature of the relationship between SVAS and WS in terms of mutational basis is less clear. In this study, we present the results of deletion mapping in classic WS and isolated SVAS patients. RAG mouse cells were hybridized with lymphoblastoid cells of WS or SVAS patients to allow their deletion breakpoints to be more finely mapped. Hybrids containing the deleted chromosome 7 from each case were screened for the presence of D7S1816, 5C19R, D7S489C, D7S489B, *ELN*, *LIMK1*, D7S2472, D7S1870, D7S489A, *HIP1* and D7S2518 in the order from centromere to telomere. Our results show that the proximal deletion breakpoint was between 5C19R and D7S489C and the distal deletion breakpoint was mapped between D7S489A and HIP1 in WS patients. On the contrary, the deletion in isolated SVAS patients was confined within the gene *ELN*. The deletion size in our WS patients is greater than that in any published data. We believe that these results would make the genotype-phenotype matching possible through cloning the genes within the deleted region and, therefore, provide valuable informations to fully understand the WS and SVAS pathogenesis.

F107 신장 투명세포육종 세포주 (CCSK-2)의 제작 및 특성분석

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신장의 투명세포육종은 소아 신장종양의 약 4-5%를 차지하는 매우 드물고 악성도가 심한 종양으로서 최근 연구결과 병리학적으로 윌름씨 종양과는 상이한 특징을 갖는 것으로 밝혀졌으며, 성인에서는 발생률이 극히 낮고 대부분 12개월에서 36개월의 소아에서 호발하는 질환이다. 본 연구에서는 19세 남성환자의 신장 종양조직으로부터 신장의 투명세포 육종세포주(CCSK-2) 개발을 시도하였다. CCSK-2는 생체외 MEM 배지에서 약 48시간의 세포배가 시간을 나타냈으며, 면역화학염색에서 vimentin에 강한 양성을 보였으나 desmin, smooth muscle actin, S-100, cytokeratin에 대해서는 음성인 것으로 관찰되었다. 이러한 결과는 확립된 세포주가 투명세포로부터 유래된 육종세포라는 사실을 강하게 시사해주고 있다. 또한 CCSK-2에서는 E-cadherin, KAI-1 및 종양억제 유전자인 p53의 발현이 억제되었고, VHL, WT-1 및 DCC는 정상 신세포주와 유사한 발현양상을 나타내었다. 향후 CCSK-2 세포주는 신장의 투명세포육종의 분자생물학적 특성 연구 및 새로운 항암성 약제 개발에 유용한 재로로서 사용될 것으로 기대된다.

F108 Sequence analysis of variations in the 5'-nontranslated region of the cardio/non-cardiovirulent Coxsackievirus B3 (CVB3) isolated in Korea

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The 5'-nontranslated region (NTR) of enteroviruses contains an internal ribosome entry site (IRES), which facilitates translation initiation of the viral open reading frame in a 5'- (m7GpppN) cap-independent manner, and cis-acting signals for positive-strand RNA replication. For several enteroviruses including Coxsackievirus B3 (CVB3), the 5'-NTR has been shown to determine the virulence phenotype. Particularly, the cardiovirulence of CVB3 has been known to be greatly influenced by the secondary structure of the 5'-NTR. We have analyzed the nucleotide sequences and compared the secondary structures of 5'-NTR of 14 non-cardiovirulent isolates and one cardiovirulent